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

Wilms tumours in children

An information guide for parents, carers and families

The purpose of this guide is to give information about Wilms tumours 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 Wilms tumours?

Wilms tumours are the most common type of kidney (renal) cancer in children. About 90 children in the UK are diagnosed with a Wilms tumour each year. It most often affects children under the age of seven.

There are two kidneys located on either side of the spine at the bottom of the rib cage. They clean the blood by removing excess fluids and waste products, which are then removed from the body through urine.

Wilms tumour was named after Dr Max Wilms, who first described it. The other name for this tumour is nephroblastoma. It is thought to develop from immature cells in the embryo. These cells are involved in the development of the child's kidneys while they are in the womb.

The cells usually disappear at birth, but in many children with Wilms tumour, clusters of these kidneys cells, called nephrogenic rests, can still be found.

Causes of Wilms tumours

It is not known what causes Wilms tumours. Very rarely, people who develop a Wilms tumour have other specific conditions which are present at birth (congenital malformations). These include the lack of an iris in the eye (aniridia), abnormalities of the genitals, and a condition where one side of the body is slightly larger than the other (hemihypertrophy). A family history is present in only 1-2% of cases and therefore it is unlikely to be passed down through family members. It is important to remember that nothing you have done has caused your child's cancer.

Signs and symptoms

The most common symptom is a swollen abdomen, which is usually painless. Sometimes, a parent or carer may feel a lump in the abdomen which can be quite large. Occasionally, the tumour may bleed slightly and this can irritate the kidney and may be painful. There may be blood in your child's urine, or their blood pressure may be raised. The child may feel tired, also have a high temperature (fever), upset stomach, weight loss or a lack of appetite.

How it is diagnosed

Your child may need a variety of tests and investigations to diagnose a Wilms tumour. These will be explained to you by your child's medical team.

An ultrasound scan of the tummy (abdomen) is usually the first thing that is done. This will be followed by an MRI and/or CT scan of the tummy and chest. These scans help doctors to identify exactly where the tumour is and whether it has spread beyond the kidney. Urine and blood samples will also be taken to check how well your child's kidneys are working. Some children will go on to have a biopsy, where a sample of tissue is taken from the tumour to confirm the diagnosis. This will depend on the medical team's assessment of your child along with results of scans and other tests.

Staging

The stage of a cancer is a term used to describe its size and whether it has spread beyond the kidney. The treatment your child receives depends on the stage of the disease. In the case of a Wilms tumour, the stage is finalised after surgery to remove the tumour. As most Wilms tumour patients receive chemotherapy before surgery, you may not know the exact stage of your child's tumour straight away. An exception is that babies under six months old usually have surgery straight away.

The staging system commonly used:

- **Stage 1** - the tumour is only affecting the kidney and has not begun to spread. It can be completely removed with surgery.
- **Stage 2** - the tumour has begun to spread beyond the kidney to nearby structures, but it's still possible to remove it completely with surgery.
- **Stage 3** - the tumour has spread beyond the kidney, either because the tumour has burst before (or during) the operation, has spread to lymph glands (nodes) in the tummy (abdomen) or has not been completely removed by surgery.
- **Stage 4** - the tumour has spread to other parts of the body such as the lungs or liver. Tumours in other parts of the body are known as metastases
- **Stage 5** - there are tumours in both kidneys (bilateral Wilms tumour).

Treatment

The main treatment for Wilms tumour is chemotherapy and surgery. Some children will also need radiotherapy. Your child's treatment will depend on the type and stage of the disease. Your child's doctor will discuss the treatment options with you.

Surgery

All children with Wilms tumour will have surgery. In a few cases, this may also involve taking a small sample of cells from the tumour to confirm the diagnosis. This is called a biopsy and is usually done under a general anaesthetic. Apart from very young children (under six months), most children will receive chemotherapy before having an operation to remove the entire tumour. The operation usually involves removing the whole of the affected kidney (nephrectomy). Most people can live normally with only one kidney remaining.

Wilms tumours can be divided into risk groups based on what they look like under the microscope. This tells doctors how they are likely to behave. Treatment following surgery depends on these risk groups, known as **low**, **standard** (or **intermediate**), and **high**.

Most tumours are in the 'standard risk' group. 'Low risk' tumours require less treatment than 'standard risk' tumours.

There are two types of high-risk Wilms tumour - anaplastic and blastemal - which require more intensive (stronger) chemotherapy:

- **Anaplastic Wilms tumour**
  About 5-10% of Wilms tumours have an appearance called anaplasia, which means the cells look very disorganised under a microscope.

- **Blastemal Wilms tumour**
  This group of high-risk tumours cannot be identified by looking at the biopsy because they occur when a particular type of early kidney cell survives the pre-surgery chemotherapy. These cells are known as blastemal cells. Tumours where most of these cells survive chemotherapy are called 'blastemal type' tumours.

Other kidney tumours

Other, less common types of kidney tumours may occur in children. These are usually only recognised after surgery to obtain a tumour sample. 'Clear cell sarcoma' and 'malignant rhabdoid tumour' of the kidney are two types of cancerous tumours, each with their own treatment recommendations.

Congenital mesoblastic nephroma is a non-cancerous (benign) tumour that occurs in very young children; this type of tumour usually only needs surgery and no other treatment.

Chemotherapy

Chemotherapy is the use of anti-cancer (cytotoxic) drugs to destroy cancer cells. It is usually given as an injection or drip into a vein (intravenously). Chemotherapy is given before surgery (pre-operative chemotherapy) to shrink the tumour and make it easier to remove.

By looking at what the tumour cells look like under the microscope and how far the cancer has spread, doctors can decide whether additional chemotherapy should be given to help reduce the risk of the cancer coming back.

The drugs used and the length of the chemotherapy depends on the stage and risk group of the tumour.

Radiotherapy

Radiotherapy treats cancer by using high-energy rays to destroy the cancer cells.

Not all children with Wilms tumour need radiotherapy. For those who do, the area to be treated depends on the stage of the tumour at diagnosis. Some children have radiotherapy to the area around the affected kidney or, less commonly, to the whole abdomen. If the tumour has spread to the lungs, then lung radiotherapy may be needed but this depends on how well the cancer responds to initial chemotherapy.

Radiotherapy is occasionally used to shrink tumours that are too large to remove surgically with the aim of allowing an operation to be done. Radiotherapy may be used when tumours have spread elsewhere in the body.

Treatment for bilateral Wilms tumour

In about 1 in 20 cases, Wilms tumour affects both kidneys. Treatment usually involves surgery to both. The aim of treatment is to remove as much of the cancer as possible, while leaving as much healthy kidney as possible. Chemotherapy is always given. Sometimes radiotherapy is needed as well.

Side effects during treatment

Treatment for Wilms tumour 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
- a sore mouth and tummy
- low blood count leading to an increased risk of infection and bruising and bleeding (sometimes blood and/or platelet transfusions are needed)
- diarrhoea or constipation

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

Relapse

About 90% of children with Wilms tumour are successfully treated. If the cancer comes back, a different course of treatment can be given. 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

Wilms tumour 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 Wilms tumour 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

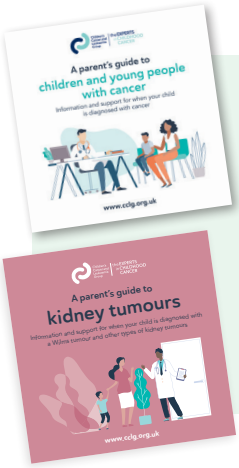
Once treatment has finished, the doctors will monitor your child closely with regular appointments to be sure that the cancer has not come back. Very few children have long-term kidney problems. After a while, you will not need to visit the clinic so often.

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 following CCLG booklets:

'A parent's guide to children and young people with cancer'

'A parents guide to kidney tumours'

are 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

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

This edition: July 2022
Next review date: July 2025

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With thanks to Dr Tanzina Chowdhury (Consultant Paediatric Oncologist, GOSH London), Dr Sucheta Vaidya (Consultant Paediatric Oncologist, The Royal Marsden Hospital, Surrey and Chair of the CCLG Renal Tumour Special Interest Group), Prof Kathy Pritchard-Jones, (Professor of Paediatric Oncology, GOSH and UCL Great Ormond Street Institute of Child Health) and Mrs Reem Al-Saadi (Translational Research Manager at UCL Great Ormond Street Institute of Child Health, London) 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. This will cost £3 plus a standard rate message.

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.

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