Wilms’ tumour

Wilms’ tumour is a type of kidney (renal) cancer in children. About 80-85 children in the UK develop a Wilms’ tumour each year. It most often affects children under the age of seven.

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. 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’.

Wilms’ tumour

Wilms’ tumour is a type of kidney cancer that was named after Dr Max Wilms, who first described it. 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 primitive kidneys cells, called nephrogenic rests can still be found.

The kidneys

The kidneys are a pair of organs found at the back of the abdomen. They clean the blood by removing excess fluids and waste products, which are then converted into urine.

Causes

In most children, the causes of Wilms’ tumour are unknown. Very rarely, people who develop 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). In 1 in 100 people with Wilms’ tumour, another family member will also have Wilms’ tumour. However, genetic factors only account for a small number of children with kidney 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 Wilms’ tumour is diagnosed**
A variety of tests and investigations may be needed to diagnose a Wilms’ tumour. An abdominal ultrasound scan is usually the first thing that is done. This will be followed by an MRI and/or CT scan of the abdomen and chest. These scans help doctors to identify exactly where the tumour is and whether it has spread beyond the kidney. This is known as staging. Urine and blood samples will also be taken to check your child’s kidney function and general health. Most children will go on to have a biopsy, where a sample of tissue is taken from the tumour to confirm the diagnosis. 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 beyond its original site. Knowing the particular type and the stage of the cancer helps doctors decide on the most appropriate treatment. 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. A commonly-used staging system for Wilms’ tumour is described below:

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
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</thead>
<tbody>
<tr>
<td>Stage 1</td>
<td>The tumour is only affecting the kidney and has not begun to spread. It can be completely removed with surgery.</td>
</tr>
<tr>
<td>Stage 2</td>
<td>The tumour has begun to spread beyond the kidney to nearby structures, but it’s still possible to remove it completely with surgery.</td>
</tr>
<tr>
<td>Stage 3</td>
<td>The tumour has spread beyond the kidney; either because the tumour has burst before (or during) the operation, has spread to lymph glands (nodes), or has not been completely removed by surgery.</td>
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<tr>
<td>Stage 4</td>
<td>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.</td>
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<tr>
<td>Stage 5</td>
<td>There are tumours in both kidneys (bilateral Wilms’ tumour).</td>
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</table>

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

**Treatment**
Treatment is planned by specialists in children’s tumours and cancers. This is usually based in a children’s oncology unit within a main hospital, but some treatment may be given closer to home.

Treatment will depend on a number of factors including how the cells appear under the microscope (histology) and the stage of the tumour. Treatment will include chemotherapy, surgery and possibly radiotherapy.

**Surgery**
All children with Wilms’ tumour will have surgery. Initially, this may only 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 using a needle inserted through the skin.

Apart from very young children (under six months), most patients will receive chemotherapy before having a bigger 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.

After examining the whole tumour under the microscope, Wilms’ tumours can be divided into a number of risk groups based on knowledge about how these different types of tumours are likely to behave. The treatment following surgery will depend on these risk groups. The risk groups are known as LOW, STANDARD (or INTERMEDIATE), and HIGH.

The majority of tumours are in the ‘standard risk’ group. So called ‘low risk’ tumours require less treatment than standard risk tumours.

Two types of Wilms’ tumour - anaplastic and blastemal - are considered to be ‘higher risk’ than other Wilms’ tumours and 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 tumour, 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’s usually given as an injection or drip into a vein (intravenously). Chemotherapy given before surgery is called neoadjuvant or ‘pre-operative’ chemotherapy.

What doctors find out about the tumour after surgery - for example, how the cells look under the microscope and how far the cancer has spread - helps them to decide whether additional chemotherapy should be given (adjuvant chemotherapy). This is to help reduce the risk of the cancer coming back (recurring). This chemotherapy may be given as inpatient or outpatient depending on tumour staging and risk group (see above).

Radiotherapy
Radiotherapy treats cancer by using high-energy rays to destroy the cancer cells, while doing as little harm as possible to normal cells.

Not all children with Wilms’ tumour need radiotherapy. For those that do, the area to be treated depends on the stage of the tumour at diagnosis. Some children receive 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; it’s not always needed.

Radiotherapy may occasionally be used to shrink tumours that are too large to remove surgically. This will, ideally, allow an operation to be done. Radiotherapy can also 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 the 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 of treatment
Treatment for Wilms’ tumour often causes side effects. Your child’s doctor will discuss this with you before treatment starts. Many side effects are expected, can be managed effectively and are reversible. Side effects can include feeling sick (nausea) and being sick (vomiting), hair loss, bruising and bleeding, tiredness, diarrhea, and an increased risk of infection. Less common effects may include impact on the heart, kidneys, and liver.

Late side effects
A small number of children may develop late side effects, sometimes many years later. These include a possible reduction in bone growth, a change in the way the heart and lungs work, and a slight increase in their risk of developing another cancer in later life. Infertility is a possible late side effect, although this is rare.

Your child’s doctor or nurse will talk to you about any possible late side effects. These will depend on the exact treatments given.

Relapse
Most (more than 85%) of children with Wilms’ tumour are successfully treated. However, for a small number of children the cancer will come back. Usually this happens after a period of time when the tumour could not be detected. This is known as tumour relapse. There are treatments available for relapsed Wilms’ tumour.

Clinical trials
Many children have their treatment as part of a clinical research trial. Trials aim to improve our understanding of the best way to treat an illness. The trial may include comparing the standard treatment with a new or modified version. It may just involve collecting patient and tumour details including response to treatment to increase specialists’ knowledge. 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 will be given plenty of time to decide if it’s right 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
Most children with Wilms’ tumour are cured. If the cancer comes back, it’s usually within the first two years. When one kidney is removed, the other will be able to work normally and can take over the work of the other kidney.

Very few children have long-term kidney problems. Your child will have regular check-ups to look for any recurrence or problems following treatment. 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
As a parent, the fact that your child has cancer is one of the worst situations you can be faced with. You may have many emotions, such as fear, guilt, sadness, anger and uncertainty. These are all normal reactions and are part of the process that many parents go through at such a difficult time.

It’s not possible to address in this factsheet all of the feelings you may have. However, the CCLG booklet ‘Children & Young People’s Cancer; A Parent’s Guide’, 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.

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.

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


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For information about the sources used to put this publication together, please contact us.

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