Wilms Tumour

A brief note for the parents.

Cancer is not an incurable disease any more in many circumstances. The key for cure is early diagnosis. However, it is not always easy to diagnose early. Some cancers do not show any signs in early stages. Survival has also improved due to medical advances in the form of better cancer drugs and better surgical treatment as well as diagnostic tests and investigations have made tremendous leaps.

It is devastating news for the parents that their child has a cancer. In Pakistan there are no support groups or organisations are available to provide solace in such difficult time. There are also difficulties in reaching the correct place and physician due to poor health system.

I have started writing about each cancer to inform parents about the cancer their child has and available treatment options in Pakistan.

If you have any questions after reading this information, you can always discuss with me through email abid@abidqazi.com and phone number +92-317-400-2444 or whatsapp +92-317-400-2444

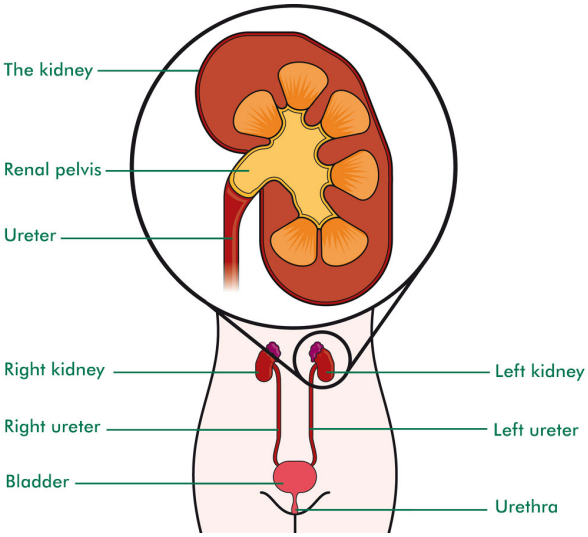
Wilms tumour

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 (blast cells) in the embryo. These cells are involved in the development of the child's kidneys while they are in the womb. That is why Wilms tumour is also known as nephroblastoma.

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. In some known associated conditions one can follow children from birth with an ultrasound scan until 5 years of age to detect early development of cancer.

The kidneys

Kidneys are a paired organ; located at the back, function to clean blood by removing toxins, waste products and excess fluid. Kidneys produce urine, which is excreted via storage in the bladder at regular intervals. Wilms tumour is the commonest childhood cancer developing from kidneys. Most children will present with this cancer before 5 years of age.



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.

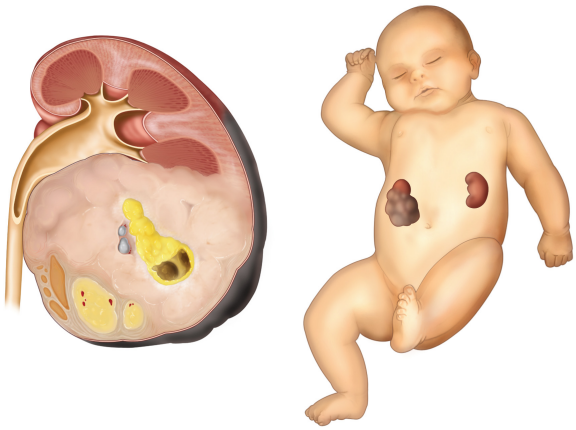
Signs and symptoms

Majority of times tumour may develop without any signs and symptoms and due to initial a slow growth parents may not be able to feel any change in the size of abdomen in their child. Later, the most common symptom is a swollen abdomen, which is still 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. A CT scan of the abdomen and chest will follow this. These scans help doctors to identify exactly where the

tumour is and whether it has spread beyond the kidney. This is known as staging of tumour. 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. You should make sure that you discuss these investigations with the treating doctor before any treatment is offered.



Staging

In Pakistan you may come across two different approaches in the treatment of this cancer. First approach is to diagnose the tumour and treat it with chemotherapy first then perform surgery to remove the involved kidney. The second approach is to remove tumour first and then give chemotherapy. In both approaches the surgeon and oncologist should be involved.

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. Staging is done differently in both approaches of treatment. A commonly-used staging system for Wilms tumour is described below:

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.

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. Depending upon which approach is used, surgery is either the first step in treatment or it is after four to six cycles of chemotherapy.

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.

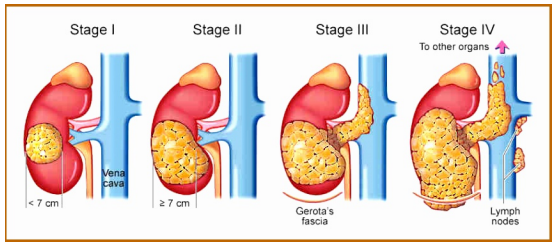
Other Kidney Tumours

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 neo-adjuvant or 'pre-operative' chemotherapy.



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), 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).

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, diarrhoea, 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.

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 world. The Children's Cancer and Leukaemia Group (CCLG) is an important organisation, which helps to produce these guidelines. SIOP and COG are the others.

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.

In case you desired, I can also get you connected to a family who have gone through the same process. This will not only give you a better insight but will also provide reassurance.

More information
can be found in our CCLG Wilms' tumour information booklet and relapse factsheet.

References

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

- Voute PA, et al. Cancer in Children: Clinical Management. 5th edition. Oxford University Press. 2005.
- Pinkerton R, et al. Evidence-based paediatric oncology. 3rd edition. Wiley-Blackwell. 2013.
- Chintagumpala M, Muscal JA. Treatment and prognosis of Wilms tumor. [online] Available from: http://www.uptodate.com/contents/ treatment-and-prognosis-of-Wilms-tumor [Accessed October 2012].

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