



Childhood Kidney Cancer

Kidney cancer rarely afflicts children and about 90 paediatric cases are diagnosed in the UK each year. About 75% of childhood kidney cancer occurs in the under-fives. The most common paediatric kidney cancer is Wilms' tumour, which make up to 95% of childhood kidney cancers. Others include the following very rare types of kidney cancer;

Clear cell sarcoma of the kidney is a type of kidney tumour that may spread to the lungs, bones, brain, and soft tissue.

Rhabdoid tumour of the kidney is a type of cancer that occurs mostly in infants and very young children. It grows and spreads quickly, often to the lungs and brain.

Neuroepithelial tumours of the kidney are rare and usually occur in adolescents. They grow and spread quickly.

Desmoplastic small round cell tumour of the kidney is a rare soft tissue sarcoma.

Cystic partially differentiated nephroblastoma is a very rare type of Wilms' tumour made up of cysts.

Renal cell carcinoma is rare in children or in adolescents younger than 15 years of age. However, it is much more common in adolescents between 15 and 19 years of age. Renal cell carcinomas can spread to the lungs, bones, liver, and lymph nodes. Renal cell carcinoma in children is sometimes associated with genetic conditions, such as **von Hippel-Lindau disease** (an inherited condition that causes abnormal growth of blood vessels), **tuberous sclerosis** (an inherited disease marked by noncancerous fatty cysts in the kidney), **neuroblastoma** and **sickle cell disease**.

Mesoblastic nephroma is a tumour of the kidney that is usually diagnosed within the first year of life and can usually be cured with surgery alone. One type of mesoblastic nephroma may appear on an ultrasound examination before birth or may occur within the first 3 months after the child is born.

Sarcomas A number of malignant tumours called sarcomas can very rarely occur in the kidney, for example **desmoplastic** small round cell tumour, **synovial** sarcoma and **anaplastic** sarcoma.



Wilms' Tumour

Wilms' tumour, or **nephroblastoma**, is a very rare type of kidney cancer which affects children. It is named after a German doctor, Dr Max Wilms (1867-1918), who first described it. Wilms' tumour afflicts about 70 children per year in the UK, and is curable in 90% of cases.

A nephroblastoma is abnormal tissue which grows on the outer part of one or both kidneys. Children with this condition are at risk of developing a type of Wilms' tumour that grows quickly. It is thought that nephroblastomas originate from specialised cells in the developing embryo known as metanephric blastema, which are involved in the development of the child's kidney while they are still in the womb. These cells usually disappear at birth, but in many children with Wilms' tumour, cells called nephrogenic rests can still be found on the kidneys.

Wilms' tumours can be categorised depending on how the tumour cells appear under the microscope as having favourable or unfavourable histology. Tumour cells with unfavourable histology look very large and not like normal kidney cells under the microscope. These cells are **anaplastic** (large and abnormal) and the cancer is less likely to be cured if there is widespread anaplasia in the tumour. However, about 95% of Wilms' tumours have favourable histology with no anaplasia and a high chance of a cure.

Causes and Risk Factors for Wilms' Tumour

The causes of Wilms' tumour in most children are unknown. However, several risk factors have been identified. Anything that increases the risk of getting a disease is called a risk factor. However, having a risk factor does not mean that you will necessarily get cancer and *vice versa*.

In a few cases, it is thought that Wilms' tumour may be part of a genetic syndrome that affects the growth and/or development of the child. A genetic syndrome is a set of symptoms or conditions that occur together and is usually caused by abnormal genes. Certain birth defects can also increase a child's risk of developing Wilms' tumour. The following genetic syndromes have been linked to Wilms' tumour:

Wilms' tumour, Aniridia, abnormal Genitourinary system and mental Retardation or WAGR syndrome This is a combination of abnormalities affecting the coloured part of the eye (iris) where the iris is partially or totally absent (aniridia), defects in the kidneys, urinary tract, penis, scrotum, testicles, clitoris or ovaries and possible mental impairment. Often children present with a milder form of WAGR with only some of these abnormalities.

Beckwith-Wiedemann syndrome Children with this condition have larger than normal internal organs, often have an enlarged tongue and sometimes one arm or leg may be bigger than the other.



Hemihypertrophy Children with this condition have one side of the body slightly larger than the other. Hemihypertrophy is linked with Wilms' tumour risk when it forms part of Beckwith-Wiedemann syndrome. However, most patients with isolated hemihypertrophy are not at increased risk of Wilms' tumour development.

Denys-Drash syndrome Boys born with this condition do not develop normal male genitalia (penis, scrotum or testicles) and can be mistaken for girls. Their kidneys do not develop properly and eventually stop working. A Wilms' tumour can grow in the damaged kidney.

There is also a slight familial link for Wilms' tumour; between 1 and 2 out of every 100 children (1-2%) with Wilms' tumour have another family member with the disease due to the inheritance of an abnormal gene from their parents.

Signs and Symptoms of Wilms' Tumour

Wilms' tumour can be very big when they are discovered due to the fact that the tumour is usually painless. In some cases the tumour can be much bigger than the kidney itself, although it is unusual for the tumour to have spread to other parts of the body from these large tumours.

These and other symptoms may be caused by Wilms' tumours. Other conditions may cause the same symptoms and a doctor should be consulted if you notice any of the following problems:

- Painless swelling or lump in the abdomen. This is the most common symptom of Wilms' tumour. Occasionally, the tumour may bleed slightly causing irritation and pain in the area surrounding the kidney.
- Blood in the urine found in 15-20% of children with Wilms' tumour.
- Raised blood pressure.
- High temperature or fever for no known reason.
- Loss of appetite.
- Weight loss.
- Upset stomach and/or vomiting.



Tests for Childhood Kidney Cancer

The following tests and investigations may be used for the detection of childhood kidney cancer, including Wilms' tumour:

Physical examination and medical history The child will undergo a physical examination of the body to check the general health of the child. It is during this examination that any lumps or swellings will be detected. A history of the child's past illnesses and treatments will also be taken.

Blood and urine samples will be taken to check the child's kidney function and general health.

Abdominal ultrasound scan and a CT scan are often done to help diagnose the tumour and to assess its growth and spread. Occasionally scans of the chest and liver may be taken to check the spread of the disease. The information obtained from these scans is used to stage the progress of the disease.

Biopsy Cells may be removed from the tumour so that they can be viewed under a microscope by a pathologist to confirm the histology of the tumour as favourable or unfavourable.

Staging of Wilms' Tumour

Wilms' tumours can be categorised into stages which describe the size and spread of the tumour. The information used to stage Wilms' tumour is obtained from ultrasound and CT scans and is used to help doctors decide on the most appropriate treatment. A commonly used staging system for Wilms' tumours is as follows:

Stage 1 The tumour is contained within 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 is still possible to remove it completely with surgery.

Stage 3 The tumour has burst before or during the operation and has spread beyond the kidney, or the tumour has spread to the lymph glands, or it has not been completely removed by surgery.

Stage 4 The tumour has spread to other parts of the body such as the lungs, liver, brain or bone to form metastases or tumours.

Stage 5 Bilateral Wilms' tumour or tumours in both kidneys.



Treatment for Wilms' Tumour

Treatment for Wilms' tumours will depend on the stage and spread of the disease, as described above, in addition to the histology of the tumour i.e. whether it has favourable or unfavourable histology. Those children with a Wilms' tumour of unfavourable histology or high risk tumour will have stronger treatment.

Surgery All children with Wilms' tumour will have surgery. In most cases, a biopsy will be taken to confirm the diagnosis and to determine the histology of the tumour as favourable or unfavourable (high risk). The tumour will be removed by surgery, usually following a course of chemotherapy to shrink the tumour. In most cases, the whole kidney is taken out (a **nephrectomy**), but occasionally only part of the kidney is removed (**partial nephrectomy**). In the case of bilateral Wilms' tumour, surgeons try to ensure that as much healthy kidney remains as possible after removal of the cancerous tissue.

Chemotherapy is usually given both before and after surgery to cause initial shrinkage and to kill off any cancer cells left behind after surgery, thereby reducing the risk of the tumour coming back or recurring. Chemotherapy is the use of drugs which are toxic to cancer cells (**cytotoxic drugs**) but relatively harmless to normal cells. These drugs stop the cancer cells from growing and can be given alone or in combination with other cytotoxic drugs, depending on the stage and histology of the tumour. Chemotherapy is usually given by means of an injection into a vein, or via a drip which requires the use of a catheter placed in a large vein, usually in the neck. Such a catheter is introduced in a small operation and usually stays throughout the course of treatment.

Radiotherapy may also be given, depending on the stage and histology of the tumour. Radiotherapy involves the use of high energy radiation to destroy cancerous cells. It can be directed to the site of the affected kidney, thereby causing the least amount of harm to surrounding normal tissue.

Chemotherapy and radiotherapy are used to treat Wilms' tumour that has spread to other organs and tissues (metastasised) i.e. stage 4 Wilms' tumour.

Chemotherapy and radiotherapy often cause side effects, which your doctor will discuss with you prior to treatment. These include feeling sick (nausea) and being sick (vomiting), diarrhoea, hair loss, tiredness, bruising and bleeding and an increased risk of infection. Additionally, some children may experience late side effects many years later, including reduced bone growth, a change in heart and lung function, infertility and a slightly increased risk of developing cancer again in later life.



Further Reading

More information about Wilms' tumours and childhood cancer in general can be found on the following websites:

<http://www.cancerbackup.org.uk> Website for Cancerbackup and Macmillan

<http://www.cancerhelp.org.uk> Website for Cancer UK

<http://www.cancer.gov/cancertopics> Website for National Cancer Institute, US National Institute of Health