



Children's
Cancer and
Leukaemia
Group

the EXPERTS
in CHILDHOOD
CANCER

A parent's guide to kidney tumours

Information and support for when your child is diagnosed
with a Wilms tumour or other types of kidney tumours



www.cclg.org.uk



Edited and reviewed by Professor Kathy Pritchard-Jones and Mrs Reem Al-Saadi (Great Ormond Street Hospital and UCL Great Ormond Street Institute of Child Health); Dr Sucheta Vaidya (Royal Marsden Hospital), on behalf of the CCLG Renal Tumours Special Interest Group; Angela Polanco (founder of Bethany's Wish); and the CCLG Information Advisory Group, comprising multiprofessional experts in the field of children's cancer.

This version is published in conjunction with Bethany's Wish, a Special Named Fund at CCLG, dedicated to Bethany Polanco, who sadly lost her life to Wilms tumour, a rare childhood cancer of the kidney. For more information, please visit www.cclg.org.uk/BethanysWish

CCLG makes every effort to ensure that information provided is accurate and up-to-date at the time of printing. We do not accept responsibility for information provided by third parties, including those referred to or signposted to in this publication. Information in the publication should be used to supplement appropriate professional or other advice specific to your circumstances. For information about the sources used to put this publication together, please contact us.

Publication of this booklet was funded by CCLG.

© 2022 CCLG

This edition: March 2022

Next review date: March 2025



About this guide

Although many children with cancer can be cured, it is devastating to hear that your child has cancer. This booklet provides general information about kidney cancer (renal tumours) that occur in children.

It is designed to provide a reference to the information you have been given already, or will be given in the future, and will act as a reminder of discussions you have had with your child's treatment team. Whilst this information may answer some of your questions, your child's specialist doctor and/or nurse will give you more detailed information, and your child's individual case should always be discussed with the team of healthcare professionals caring for your child.

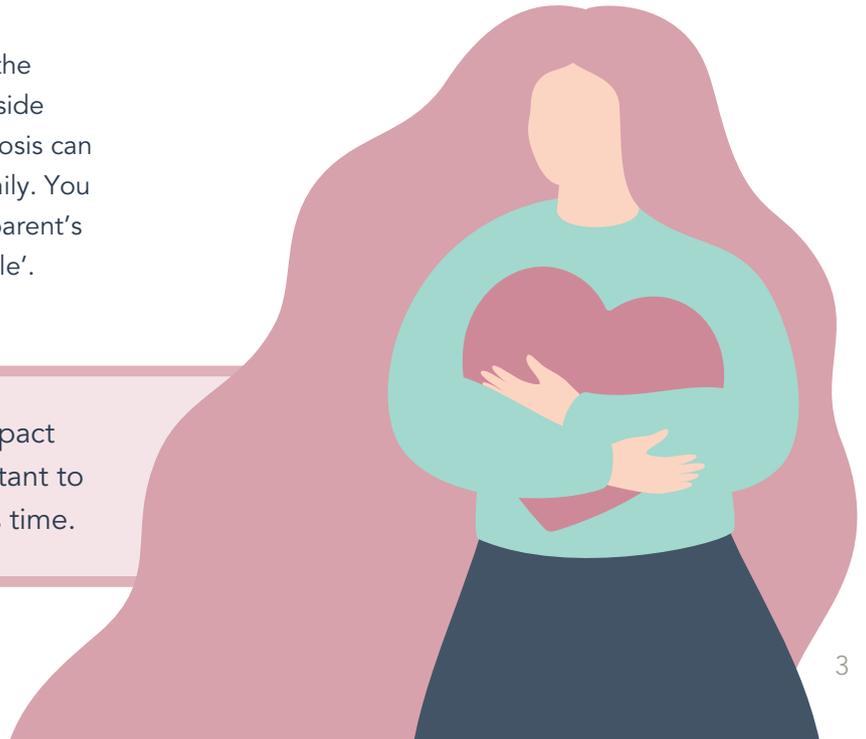
This booklet contains information about the different types of childhood kidney cancer, the treatments that are used and their possible side effects. It also discusses how a cancer diagnosis can affect you, your child and the rest of the family. You may also find helpful the CCLG booklet 'A parent's guide to cancer in children and young people'.

Cancer and its treatment has a big impact on you and your child's life. It's important to have support to help you through this time.



For more detailed information on caring for yourself and others, please read our accompanying booklet 'A parent's guide to cancer in children and young people'.

www.cclg.org.uk/publications



Contents

Beginning the journey	5
About childhood kidney tumours	9
• How common are kidney tumours in children?	10
• Types of childhood kidney tumour	11
• What causes kidney tumours?	12
• Signs and symptoms of kidney cancer	13
• Urgent referrals	13
Diagnosis	15
• How cancer is diagnosed	17
• Tests and scans	17
• Before treatment starts	18
• Commonly used staging for Wilms tumour	19
Starting treatment	21
• Your child's treatment and care	22
• What your child's cancer care might look like	23
• Types of treatment	24
• Side effects of treatment	25
• Treatment options for Wilms tumours	26
• Treatment options for other childhood kidney tumours	28
• Other very rare types of kidney cancer in children	31
• Taking part in clinical trials	31
• Giving your consent	32
• Randomisation	32
• Safety	32
After treatment	21
• Living with a single kidney	34
• Keeping your child's single kidney healthy	34
• Relapsed Wilms tumour	35
• If treatment doesn't work	35
Caring for yourself and other family members	37
Tumour cell types and glossary	43
Help and support	49

Beginning the journey

When you are told that your child has a kidney tumour, it can feel as if your life has turned upside down overnight.

You may be feeling numb, scared, not believing that any of this is really happening, angry that this is happening to your child and maybe feeling guilty that your child's cancer is because of something you have or haven't done, even though this isn't true. All of these feelings are completely normal and many parents say that they felt the same.

Since your child's diagnosis, you may have met many new people, heard a lot of unfamiliar medical terms and your child may have undergone a series of tests. This can feel very overwhelming and daunting as a parent. Don't worry, hospital staff fully understand that it takes time for you to digest what is happening and what the next steps might be. They are there to help you through this difficult time with information and reassurance.

Many parents cannot think of any questions to ask during a hospital consultation but think of all sorts of things as soon as they get home. It is a good idea to write down questions as soon as you think of them so that you can discuss them at the next opportunity.

When a child is diagnosed with cancer, it has a huge impact on your whole family. Your child's routine is likely to change with hospital stays and regular appointments and this can feel overwhelming for you, your child and the rest of your family.

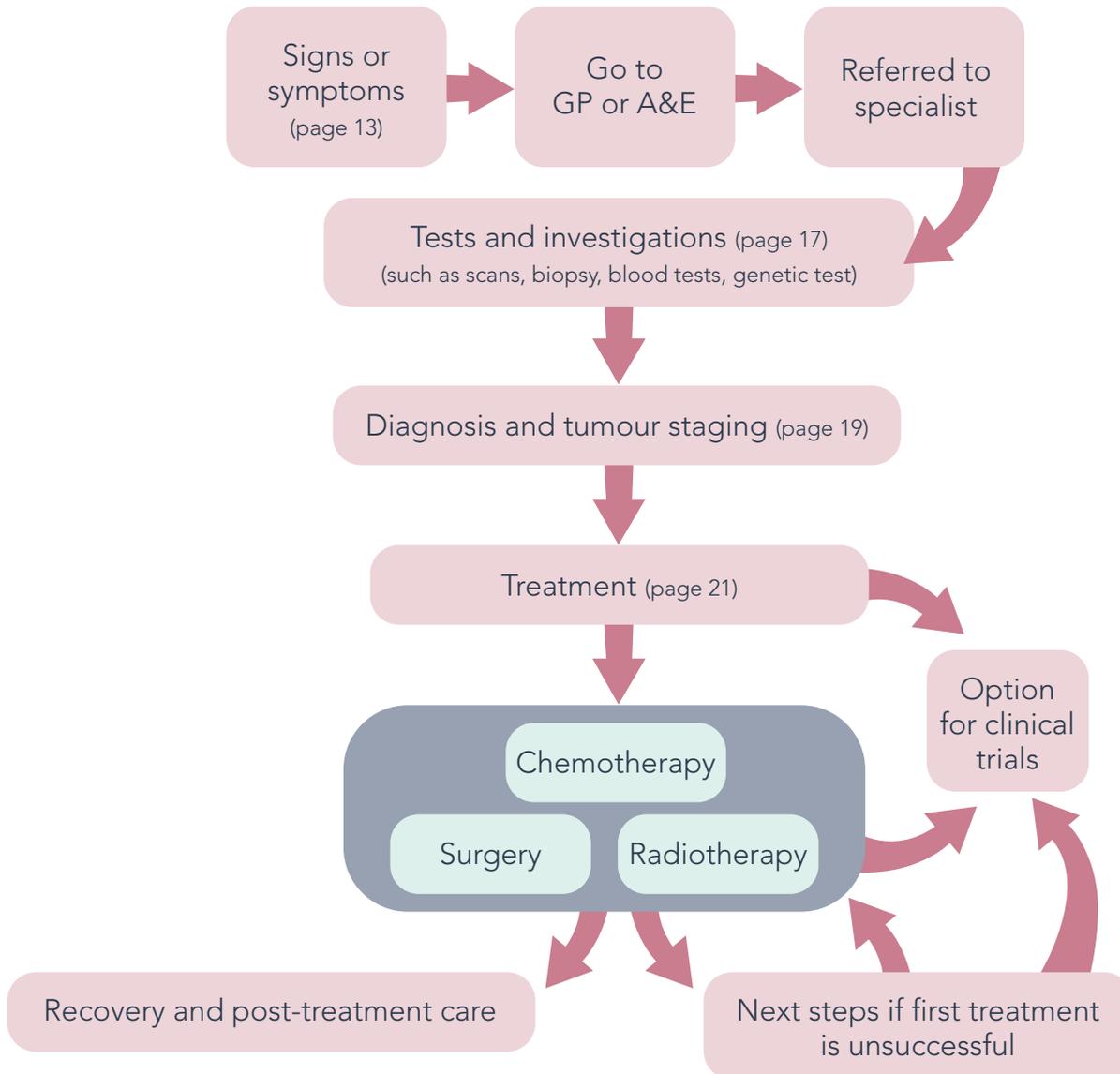
There are many support organisations who can help you through this time and some of these organisations are listed at the end of this guide, but you should also discuss your feelings with the team looking after your child.

TOP TIP

Write down your questions at the back of this guide and fill in the answers during your clinic appointments



Your child's healthcare journey





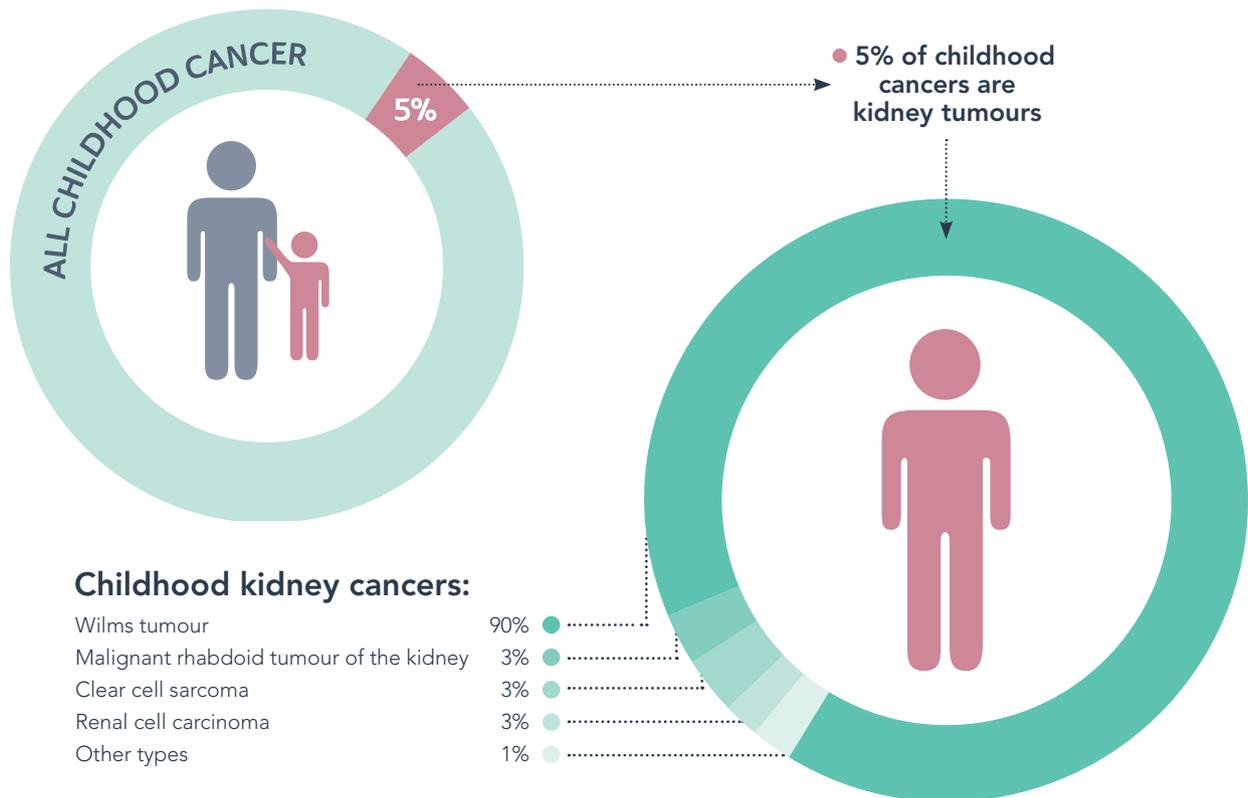
About childhood kidney tumours

Childhood kidney cancers, also called renal tumours, are types of cancer that develop in the tissues of the kidneys.

There are two kidneys located on either side of the spine at the bottom of the rib cage. The kidneys' main function is to filter and clean the blood by removing excess fluids and waste products, which are then removed from the body through urine. Other key functions include keeping the body's salts in balance and controlling red blood cell production and blood pressure.

How common are kidney tumours in children?

In the UK, almost 1,700 children are diagnosed with cancer each year. About 5% of these, or around 80-85 children, are diagnosed with kidney cancer.



Types of childhood kidney tumour

Wilms tumour (sometimes called 'nephroblastoma') is the most common type of kidney cancer in children. More than 90% of kidney cancers in children are Wilms tumours.

Wilms tumour is almost always diagnosed in children under seven years old, and in 90% of cases it affects just one of the two kidneys (unilateral disease).

Most cases of unilateral Wilms tumour occur at around three years of age, and it is slightly more common in girls than boys. Wilms tumour is an embryonic kidney cancer thought to develop from immature cells in the baby/or embryo in the womb. These cells are involved in the development of the kidneys while a child is in the womb, and usually disappear at birth. However, in many children with Wilms tumour, clusters of these cells can still be found.

The outlook, or prognosis, for children with Wilms tumour is very good. In Europe and North America, 90% of children diagnosed with Wilms tumour survive for five years or more.



It is not known what causes Wilms tumour but several risk factors have been identified. The development of Wilms tumour is thought to be partly due to a genetic cause, and a number of genetic abnormalities and malformation syndromes are associated with developing Wilms tumour. A family history is present in only 1–2% of cases, and therefore it is unlikely to be hereditary.

Malignant rhabdoid tumour of the kidney (MRTK) and **clear cell sarcoma of the kidney (CCSK)** each account for 2-3% of cases. Both of these cancers were initially thought to be higher risk variants of Wilms tumour but now it is known that they are different cancers.

Primitive neuroectodermal tumour (PNET) of the kidney accounts for less than 1% of new cases.

Renal cell carcinoma is common in adults but is hardly ever seen in children. Fewer than 3% of all childhood kidney tumours are renal cell carcinomas.

If this type of cancer occurs in children, it has a different 'subtype' and has different features to the common adult renal cell carcinoma.

Other types of childhood kidney cancer, including **rhabdomyosarcoma of the kidney**, **desmoplastic small round cell tumours of the kidney**, **primary renal synovial sarcoma**, and **anaplastic sarcoma of the kidney**, which account for less than 1% of cases between them.

In addition to malignant tumours, several types of benign (non-cancerous) kidney tumours also occur in childhood. The most common of these is **mesoblastic nephroma**. This is usually a benign tumour that is often diagnosed during pregnancy, when it is seen on an ultrasound scan. It is usually treated with surgery only.

Nephrogenic rests are immature cells considered to be potential precursors for Wilms tumour. If they are visible as a lump on scans, they may need chemotherapy and surgery.

What causes kidney tumours?

As with most childhood cancers, the causes of most childhood kidney cancers are unknown. It is not infectious and cannot be passed on to other people.

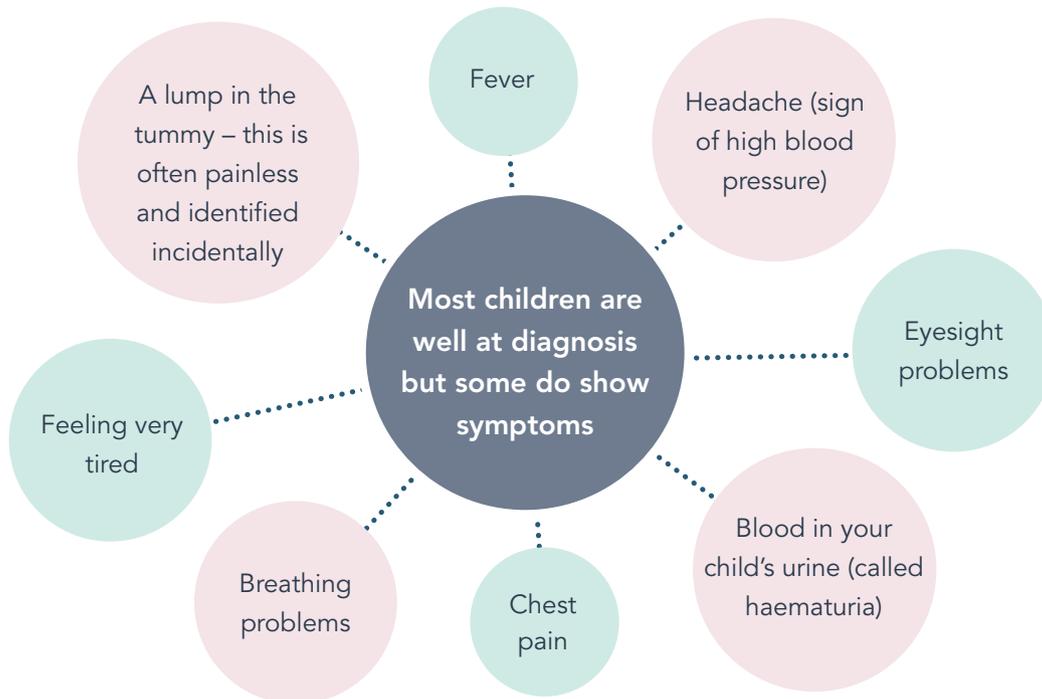
There are certain factors (risk factors) that increase the chance of kidney cancer occurring in some children, such as inherited (genetic) factors. These genetic risk factors are different depending on the type of kidney cancer. Genetic factors only account for a small number of children with kidney cancer.

Signs and symptoms of kidney cancer

Many children with a kidney tumour have little in the way of symptoms. The first sign is usually a lump in the tummy followed by blood in the child's urine. Cancer is rarely suspected if only vague symptoms are present. If cancer has spread to the lungs or liver, then this may cause a cough, trouble with breathing, or pain.

Urgent referrals

The National Institute for Health and Clinical Excellence (NICE) referral guidelines for suspected cancer recommend very urgent referral (for an appointment within 48 hours) for specialist assessment if a child has a swollen abdomen or blood in their urine.





Diagnosis

If cancer is suspected, your child will be referred by your GP or A&E staff to your nearest children's hospital which has a specialist children's cancer (called paediatric oncology) centre. These centres are located in a network of hospitals across the UK and Ireland with expertise in managing and treating all childhood cancers, including kidney cancer.

1 Aberdeen:
Royal Aberdeen Children's Hospital

2 Edinburgh:
Royal Hospital for Children and Young People

3 Glasgow:
Royal Hospital for Children

4 Newcastle-Upon-Tyne:
Great North Children's Hospital,
Royal Victoria Infirmary

5 Leeds:
Leeds Children's Hospital,
Leeds General Infirmary

6 Sheffield:
Sheffield Children's Hospital

7 Nottingham/Leicester:
East Midlands Integrated Service at
Queen's Medical Centre, Nottingham
and Leicester Royal Infirmary

8 Cambridge:
Addenbrooke's Hospital

9/10 London:
Great Ormond Street Hospital
for Children and University College
Hospital London

11 Sutton:
Royal Marsden Hospital

12 Southampton:
Southampton Children's Hospital,
Southampton General Hospital

13 Bristol:
Bristol Royal Hospital for Children

14 Cardiff:
Noah's Ark Children's Hospital for Wales

15 Oxford:
Oxford Children's Hospital,
John Radcliffe Hospital

16 Birmingham:
Birmingham Children's Hospital

17 Liverpool:
Alder Hey Children's Hospital

18 Manchester:
Royal Manchester Children's Hospital

19 Dublin:
Our Lady's Children's Hospital, Crumlin

20 Belfast:
Royal Belfast Hospital for Sick Children



How cancer is diagnosed

Kidney cancer is diagnosed through a combination of tests on the kidneys and blood. The first of these is a physical examination to check general signs of health and look for signs of disease, such as lumps or anything else that seems unusual. This is coupled with questions about the child's medical history.

If kidney cancer is suspected, the next test is an ultrasound examination that can be done without anaesthetic. If this confirms a lump in the kidney, your child will need CT and/or MRI scans to check the appearance of the kidneys, and to see if there are any signs of cancer spreading to the liver or chest – this is called **tumour staging**. A general anaesthetic or sedation is often needed to perform these tests. An x-ray may also be taken.

The diagnosis can be made with the aid of imaging such as MRI or CT scan but for most children a biopsy is not needed. In older children (from seven years old onwards) and in some younger children with certain imaging features and test results, a biopsy is recommended. Doctors will also check how the kidneys are working (**kidney function**) by testing a urine sample to check for protein and blood, and by measuring the levels of salts in the blood.

Scans will also show if one or both kidneys are affected, and whether there is a kidney abnormality (1-2% of children with Wilms tumour have a horseshoe-shaped kidney).

Tests and scans

Tumour biopsy

A small piece of tumour is often taken for examination. This test is known as a biopsy. It involves an operation where your child has a general anaesthetic and a piece of the tumour is taken out through a small cut (incision) or needle puncture in the skin.

A series of tests may be carried out on the cells in this biopsy to find out more about the biology of the tumour. Knowing about tumour biology provides information that is used in deciding the best treatment for your child.

Blood tests

Blood for testing may be taken from a vein in your child's arm or by a finger prick. You are probably familiar with both of these procedures.

X-rays

X-ray images may be taken to investigate the location of the tumour and whether the tumour has spread.

CT (computerised tomography) scans

The CT scanner takes multiple x-ray images and these are converted by a computer to form a 3D view of either the whole body or of the part of the body under examination.

Ultrasound scans

Similar to an ultrasound scan during pregnancy, the sound waves produced by the scanner bounce from solid organs inside the body and are recorded on a screen. The doctors can see the outlines or shadows of normal organs and of any tumour inside the body.

MRI (magnetic resonance imaging) scans

An MRI scan relies on magnetism and is a very safe procedure as no radiation is used. It takes longer than a CT scan and is quite noisy. There are no known side effects to this type of scan.

Apart from the need for an injection of 'contrast' during some CT scans, none of these investigations are painful to your child, but it is appreciated that they may feel unsettled or frightened. Some of the scans require that your child remains still for quite some time and to assist in this sedation may be given. An anaesthetic may be needed for some children to have some of the tests.

It may take several days for any tests to be completed and the results analysed. Undergoing these tests and waiting for results can be a stressful time and lead to anxiety. However, exact assessment of the extent of your child's disease before beginning treatment is very important. The results obtained will inform the type and length of treatment that will be given to your child.

Before treatment starts

The UK is an active member of the International Society for Paediatric Oncology Renal Tumour Study Group (SIOP-RTSG: www.siop-rtsg.org). This is a group of doctors from several countries who are experts in the specialist areas that are needed to treat children with kidney cancers. They work together to decide on the best treatments currently available and to design clinical trials and studies that aim to improve survival for all children with kidney tumours and to test new treatments or better ways of giving treatments for some tumour types. In the UK, treatments for childhood kidney cancers are based on results of clinical trials performed by the SIOP Renal Tumours Study Group.

Since 2019, the current clinical study of the SIOP-RTSG is called 'UMBRELLA'. This is an observational study that includes all types of childhood kidney tumours. By registering on the study, you agree for clinical information, copies of your child's scans and samples of your child's tumour, blood and urine to be used in research. The main research question is to decide if tumour biomarkers (genetic changes in the tumour) can be used to change treatments in the future. The course of treatment recommended for your child will be discussed with a **multi-disciplinary team (MDT)** at your main hospital. The MDT is a group of doctors and other health professionals with expertise in

childhood cancer, who together discuss the best course of treatment for their patients. This team will include oncologists, surgeons, pathologists, radiologists and specialist nurses. Decisions about how to treat your child's tumour will be determined by a number of

factors including the specific cells that make up the tumour, the stage of the cancer, the type and size of the tumour, the age of your child, whether the tumour can be removed by surgery, the presence of genetic abnormalities, and whether the cancer is newly diagnosed or has recurred.

Commonly used staging for Wilms tumour

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 is 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).



Starting treatment

Once your child has a confirmed diagnosis of kidney cancer, the medical team will be keen to get your child started on treatment as soon as possible at your nearest specialist Principal Treatment Centre (PTC) for children's cancer. This means that your child will get highly specialised care and may need to stay in hospital for a while so that they can be monitored when starting treatment.

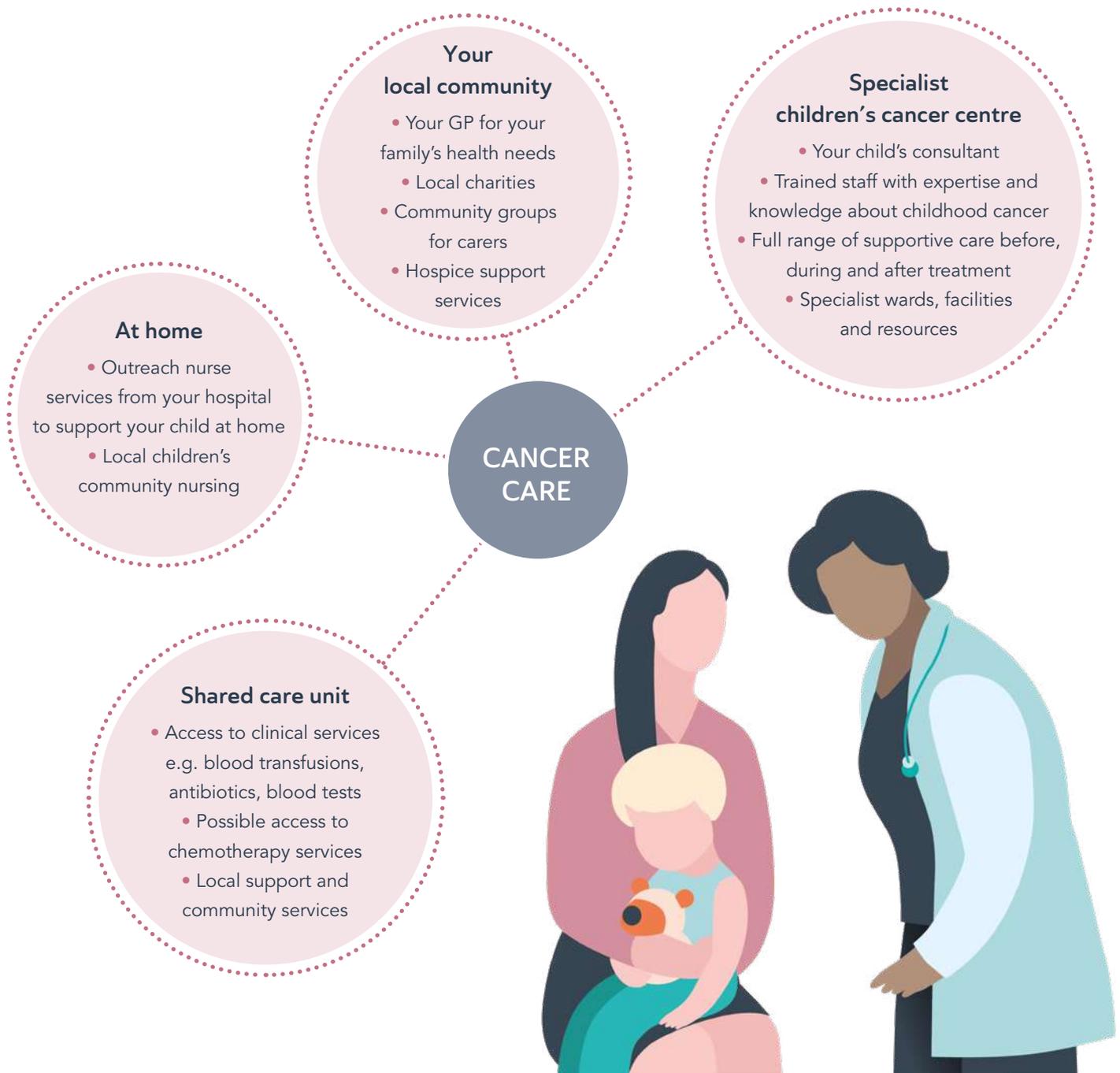
Over time, your child may also receive some treatment at your local hospital. This is called 'shared care' and allocated local hospitals are known as Paediatric Oncology Shared Care Units (POSCU). If your child has any shared care, the decisions about their treatment will still be made by your child's consultant at the main cancer unit but it just means that it is easier for families as they will need to travel less.

A lot of different professional staff will be involved in looking after your child, both at home and while they are in hospital. A team of specialists called the 'multi-disciplinary team' (MDT) will meet regularly to discuss your child's cancer, treatment options, clinical trials, and any areas in which your child may need support.

Your child's treatment and care will be managed by:

- **Consultant paediatric oncologist** – a doctor who specialises in treating all children with cancer
- **Consultant clinical oncologist** – a doctor who specialises in using radiotherapy to treat cancer patients
- **Clinical nurse specialist** – a nurse who specialises in caring and supporting children with cancer

What your child's cancer care might look like:



Types of treatment

Surgery

Your child's tumour will be removed during an operation if it is possible and safe to do so. In some children, surgery may be involved at a later stage of treatment. This is often after several cycles of chemotherapy have been given to shrink the tumour so that it can be more easily (and therefore more safely) removed by the surgeon. The extent of surgery differs for each child and will depend on factors such as the location of the original tumour and involvement of surrounding organs or structures.

If your child has received chemotherapy before surgery, then it is important to make sure that your child has made a full recovery before surgery takes place. This will be discussed with you.

Chemotherapy

Chemotherapy is the use of drugs to destroy cancer cells. They can be given in different ways but the most common way is intravenously – into a vein – whether through injections, cannula, or a line such as a central line, implantable port or PICC line. Chemotherapy is usually given as a combination of multiple different drugs. Once the diagnosis is confirmed, the results from the tests will help your child's doctor to decide on the best treatment regimen often called 'protocol'. The suggested treatment will be discussed fully with you.

Radiotherapy

Radiotherapy treats cancer by using high-energy rays to destroy cancer cells in a specific part of the body. Radiotherapy is focused on the area where the primary tumour was removed at the time of surgery. It is used to try and kill tumour cells that can sometimes remain after surgery.

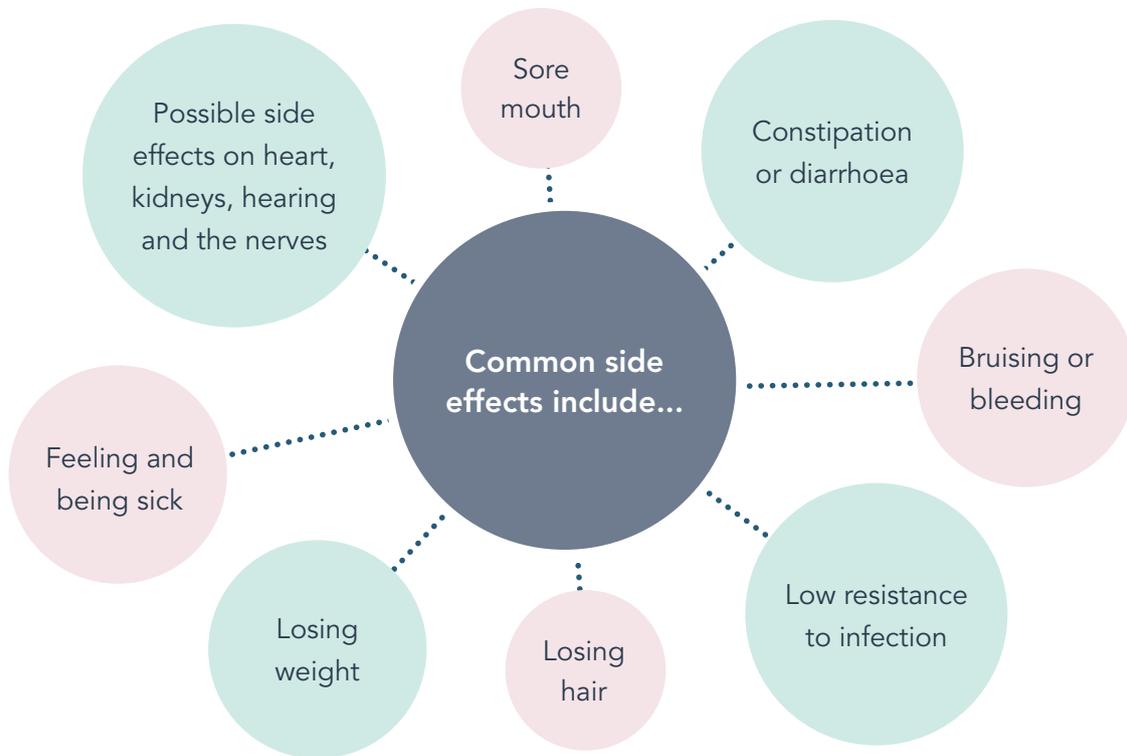
Radiotherapy is painless and the machine does not touch your child – it is like having an ordinary x-ray. The total radiation dose is spread out over time and often involves treatment every day for 3-5 weeks, usually excluding weekends. In some situations, the radiotherapy course can be shorter or longer and your child's doctor will discuss this with you.

Radiotherapy requires careful preparation and planning to decide where in your child's body to treat and the best way to give the radiotherapy, and can involve several steps. Some young children may struggle to lie still for radiotherapy so sometimes need general anaesthetic each day for radiotherapy planning and treatment. This will be discussed with you by your child's clinical team.

Side effects of treatment

There are a number of possible side effects from treatment. These are usually short term, lasting only while the treatment is given and depend on the drugs used. The risks of any side effects of treatment will be carefully explained to you. The chemotherapy drugs most often used to treat

Wilms tumour are vincristine, actinomycin D and sometimes doxorubicin. All can have side effects that will be carefully explained. A serious side effect of actinomycin D is liver toxicity, but this occurs only rarely. A rare late effect of doxorubicin may cause heart problems which will be carefully explained.



Treatment options for Wilms tumours

Treatment will depend on the size of the tumour, if it has spread outside the kidney (**stage**) and the appearance of cells under the microscope. (**histology**).

In the UK and Europe, treatment for Wilms tumour is based on guidelines developed by the SIOP-RTSG. Treatment typically consists of first treating your child with chemotherapy to shrink the tumour and make surgery easier, followed by an operation to remove the kidney containing the tumour (nephrectomy).

The type of chemotherapy needed after surgery and whether radiotherapy is also needed depends on the tumour histology and stage. An overview of the treatment pathway for Wilms tumour in the UK is shown below.

The number of drugs involved and length of chemotherapy before surgery depends on whether the disease has spread outside the kidney. After surgery, the tissue removed by surgery is examined under a microscope to assess the effect of any pre-operative (given before surgery) chemotherapy and determine what treatment is needed after surgery. Based on the histology, the tumour is classified into three risk groups - low, intermediate and high.

- **Low:** very few tumours fall into this group and, for some, a short course of treatment is enough
- **Intermediate:** most Wilms tumours fall into this risk group
- **High-risk:** two type of Wilms tumour called anaplastic and blastermal type are high-risk and need stronger chemotherapy

These risk groups help doctors to decide the best course of treatment. Some patients with higher risk tumours will go on to receive radiotherapy to the tumour, but also to the chest if there is evidence of the disease having spread at the time of diagnosis.

Pre-operation	Operation	Post-operation
No evidence of tumour spread beyond the kidney (localised disease). Chemotherapy for 4-5 weeks with 2 drugs.	Removal of whole kidney (called a nephrectomy).	Between 4 and 35 week course of chemotherapy (depending on the stage and histology of the tumour).
Tumour spread outside kidney (Stage 4). Chemotherapy for 6-7 weeks with 3 drugs.	Partial nephrectomy and bilateral (not stage 4)	Radiotherapy: <ul style="list-style-type: none"> • Flank (between the ribs and the hip) only for tumours that are Stage 3 or more.
Bilateral Wilms tumour (Stage 5). Chemotherapy for 6-12 weeks with 2 drugs.		<ul style="list-style-type: none"> • Lungs only for metastases that are slow to respond or high-risk histology.

Bilateral Wilms tumour

Between 5-10% of children have Wilms tumour affecting both kidneys at the same time (**bilateral Wilms tumour**). Whilst it is still very possible to cure children with bilateral Wilms tumour, it poses additional challenges, particularly at the point of surgery.

The emphasis for these children will be to give a longer chemotherapy course before surgery in order to get the maximum shrinkage of tumours on both sides before surgery. It is often possible to spare kidney tissue at the operation by performing 'nephron-sparing surgery'. This involves a great

deal of discussion beforehand; both at your child's MDT and sometimes at the National Renal Advisory Panel (NRAP) meeting. This is run by members of the national CCLG Renal Tumour Group in the UK to help local centres decide on the best approach.

It is very rare nowadays that children will lose both kidneys and then require dialysis until a kidney transplant is available.

Each child with bilateral Wilms tumour has a personalised treatment plan, aiming for the best possible chance of curing the tumour and keeping enough kidney function in the long term.

Treatment options for other childhood kidney tumours

Around 10% of childhood kidney tumours are not Wilms tumours, and are made up of a variety of rare tumours.

Clear cell sarcoma of the kidney (CCSK)

Clear cell sarcoma of the kidney (CCSK)

makes up 2-3% of all kidney cancers in children.

CCSK affects children at about the same age as Wilms tumour (around three years of age), and usually presents with the same types of symptoms (lump in the tummy, blood in the urine). It is not easy to tell the difference between the two types of tumour on imaging scans, and diagnosis of CCSK is usually made from a biopsy of the tumour or following surgery.

CCSK occurs more commonly in boys and the cancer sometimes spreads to the bones or brain, which is unusual in Wilms tumour. CCSK does not appear to be associated with malformation syndromes, and familial cases have not been reported. Survival from this type of kidney cancer is almost as good as for Wilms tumour provided stronger treatment is given.

CCSK treatment also includes surgery, chemotherapy and radiotherapy. However, the chemotherapy is stronger and given over 34 weeks following surgery. All children will be treated with the drug doxorubicin as part of their chemotherapy regimen and most children (except if they have

a stage 1 tumour) will have post-operative radiotherapy. About one in four children with CCSK will relapse and need additional treatment.

After treatment finishes, children will continue to be monitored and assessed and will receive follow-up care well into adult life. The follow-up tests for children with CCSK will include (abdominal ultrasound scans, chest x-ray, urine tests and blood tests). If the cancer has spread to other organs such as the brain or bones when the cancer was diagnosed, additional tests may be needed and these parts of the body may also be checked during follow-up.

Malignant rhabdoid tumour of the kidney (MRTK)

Malignant rhabdoid tumour of the kidney (MRTK)

accounts for between 2 and 3% of all kidney tumours in children. While MRTK was first recognised in the kidney, it is now known to occur elsewhere in the body as well.

Children with this tumour are generally found by their doctor to have a detectable abdominal lump plus other signs, such as blood in the urine, fever, infection, high blood pressure and

anaemia. MRTK is found in younger children, and two-thirds are diagnosed under 12 months old. Boys and girls are equally affected.

This tumour has a characteristic genetic change, and nearly all MRTK tumours have a mutation in a gene called INI1 (sometimes also called SMARCB1, BAF47, or hSNF5). There are rare families that carry a mutation in this gene, and sometimes in these families more than one case of this tumour has occurred. If your child has this type of tumour, your doctor may discuss genetic screening for your family.

This tumour looks similar on imaging by ultrasound, CT or MRI to Wilms tumour, and diagnosis requires examination of a biopsy sample. In particular, the genetic alteration in the INI1 gene is characteristic of MRTK and does not occur in other cancers. Doctors can use this genetic alteration to confirm a diagnosis of MRTK.

MRTK is highly aggressive and grows rapidly, often affecting both kidneys. About two-thirds of children have advanced disease when they are diagnosed.

In the UK, MRTK is treated with intensive chemotherapy, surgery, and then chemotherapy again, in conjunction with radiotherapy. Even with such treatment, the outlook for patients with this disease is generally poor, although some children have experienced extensive responses.

New treatments for this cancer are being sought, and the best treatment option for your child would usually be to enter a clinical trial.

Renal cell carcinoma (RCC)

Renal cell carcinoma (RCC) is the most common form of kidney cancer in adults but it is rare in children younger than 15 years old. Many children with RCC have a type called 'translocation renal cell carcinoma'. This type is known to be caused by a specific genetic alteration involving a gene called TFE3 or TFEB. About 50-70% of children and young adults with RCCs have this alteration in their tumour but not in their blood (non-hereditary). Several other subtypes of renal cell carcinoma can be found in children and young adults.

The most common features found at diagnosis are pain, blood in the child's urine and a tummy lump. Fever, weight loss and lethargy may also be seen. There are several genetic syndromes associated with RCC and your child's doctor may check to see if your family might have one of these related syndromes and in some cases recommend genetic screening. Most children with RCC do well but overall prognosis is dependent on the stage of the tumour at diagnosis.

Treatment for children with RCC will be given at a specialist children's cancer centre with a treatment plan determined by a multi-disciplinary team. Surgery is the cornerstone of treatment for this type of kidney cancer. Many children with RCC require only surgery and have done well without chemotherapy, even if the disease had spread to their lymph nodes. In patients with cancer that has spread, some chemotherapy treatments, and newer types of drugs that target the abnormal proteins that result from changes in the tumour DNA, such as sunitinib, may be effective. As for other types of rare kidney cancers, your child's doctor may suggest that the best treatment option is for your child to be part of a clinical trial.

Mesoblastic nephroma

Mesoblastic nephroma is sometimes called congenital mesoblastic nephroma (CMN) because it may be present at birth but the average age of diagnosis is 3-4 months. CMN is very uncommon, with only 4-5 cases a year diagnosed in the UK. It is generally considered a benign (non-cancerous) tumour but it may behave like a cancer in certain cases.

The young children affected by mesoblastic nephroma often don't have any symptoms, and a lump is detected during a routine examination. More and more cases are being discovered during the regular ultrasound scans done during pregnancy. In these cases, there is usually no rush

for the baby to be born early, unless the tumour is causing strain on the heart because of increased blood flow through the tumour.

Some older children with mesoblastic nephroma will have a lump in their tummy, high blood pressure and blood in their urine. There are two main subtypes of mesoblastic nephroma that are only distinguished under the microscope: classic (or conventional) and cellular (or atypical). Cellular mesoblastic nephroma is typically distinguished by a characteristic genetic change involving genes called ETV6 (TEL) and NTRK3. Classic disease tends to be found in very young babies or before birth, and cellular disease tends to be found in older infants. However, some patients show a mixture of the two types. Mesoblastic nephroma is usually treated with surgery only. This treatment usually leads to excellent outcomes, with 95% survival rates. Even if the tumour has started to spread, doctors will often choose not to treat with any other therapy. The cellular subtype is known to respond to various combinations of chemotherapy.

Other very rare types of kidney cancer in children

There are other types of kidney cancer that have been identified in a limited number of children.

- **Primitive neuroectodermal tumour (PNET)** of the kidney is very rare, with only approximately 50 cases known to medicine.
- **Primary renal synovial sarcoma of the kidney** is also very rare, with fewer than 70 cases described since its characterisation in 1999.
- **Anaplastic sarcoma of the kidney** usually affects females and has a broad age range but has been documented in children.
- **Rhabdomyosarcoma of the kidney** is a rare soft tissue sarcoma usually diagnosed in children around six years of age.

Treatment will depend on the type of cancer but typically will include surgery, chemotherapy and radiotherapy. If your child has one of these rare types of kidney cancers, they will be treated in a specialist cancer centre and the MDT will determine the best treatment plan for your child. This may include participation in a clinical trial.

Taking part in clinical trials

Most children treated for kidney cancer at one of the specialist children's cancer units in the UK are offered cancer treatment within a clinical trial. These are research studies carried out to try and find new and better treatments for cancer such as a new drug or combination of treatments. By doing this through clinical trials, we can make sure that comparisons are measured consistently and reliably over time to see if one treatment is better than another. This is why improvements are made every year in treating and curing cancer in children.

Your child will receive the best possible treatment regardless of whether they are on a trial or not. If a trial is suggested, you will be given information about it and what is involved.

You will need to consent to the trial as well as to the treatment itself. Your child's consultant will be able to discuss potential clinical trials with you.



Giving your consent

Before your child is asked to take part in a clinical trial, your child's medical team will explain what the trial is aiming to achieve and the risks and benefits of taking part. Once you have considered everything you need to know, you will be asked to give your consent for your child to take part and to sign a form. There is no pressure to take part and your child will receive the best treatment available whatever you decide.

When deciding whether to take part in a trial, it can add to the stress of coping with your sick child and the bewildering range of processes associated with treatment. It may seem as though the treatment team is passing over responsibility for deciding what treatment your child should be given. However, in practice, the treatment team will only invite you to join a trial when it is considered in the best interests of your child and where both you and your child are likely to benefit.

Randomisation

For some trials, the researchers carry out a process called randomisation. This means a computer will randomly allocate your child to have a particular treatment in the trial. This is done so that each treatment group has a similar mix of children of different ages, sex and general health. It makes sure that researchers and doctors can't decide who should get which treatment to avoid bias that could skew the findings.

Safety

The safety of children in clinical trials is the top priority. All trials are approved by ethics and regulatory committees, and they are reviewed on an ongoing basis.

After treatment

When treatment finishes, your child will be given a summary of the treatment they have received and an aftercare plan involving regular follow-up checks in hospital over the next few years.

This is to make sure there are no signs of the cancer coming back (called 'relapse') and also to look for and treat any long-term effects that might happen as a result of the cancer and its treatment (called 'late effects'). These possible late effects can affect their heart and lungs, growth hormones, and they may be at a higher risk of developing cancer again in the future, so it is important that your child attends their follow-up clinic appointments.

In general, from the date treatment finishes, your child will have an ultrasound scan of their abdomen (tummy) every three months for two years and regular chest X-rays every three months for three years.

At each clinic visit, doctors will check your child's blood pressure and urine. If any of these tests are abnormal, your child's doctor may perform additional tests such as a CT scan, MRI scan or extra blood tests.

An annual check of blood pressure and protein in the urine is recommended for children with only one kidney for the rest of their lives through their family doctor (GP). Blood tests to check kidney function can be done less often.

Children who were treated with the drug doxorubicin as part of their chemotherapy will need an ultrasound scan of the heart (echocardiogram) every few years.

Living with a single kidney

It is perfectly possible to lead a completely normal life with only one kidney, but it is important that your child looks after their remaining kidney throughout their life. There are some very simple measures that can be taken to check that the kidney is working well. This will include an annual blood pressure check and urine test to look at how well the kidney is at filtering waste from the body.

Keeping your child's single kidney healthy

- Drink plenty of fluid - especially on a hot day
- Eat a healthy, balanced diet
- Exercise regularly (see below)
- Do not ignore symptoms that might be a urinary tract infection (see below)

You should not ignore symptoms that might be a urinary tract infection; for example, fever, tummy ache and a burning sensation while passing urine. If this occurs, you should see your child's GP.

Regular exercise is important. If taking part in contact sports, particularly as your child becomes older, some doctors may advise body padding to avoid injuring the single kidney. Talk to your doctors before taking up competitive sports to discuss this further.

Relapsed Wilms tumour

For most children with Wilms tumour, their first treatment is successful. However, for a small number of children, the cancer will come back. When this happens, it is usually 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.

The CCLG factsheet 'Treatment options for children with relapsed Wilms tumour' 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



If treatment doesn't work

For some children, parents are told the devastating news that their child's cancer is not curable. Your child's care will change to focus on giving the best quality of life for your child and managing their symptoms. There is a huge amount of practical and emotional support for families at this time, such as from your child's hospital, charities, and organisations.

CCLG has a range of resources to help parents to prepare and plan for the next steps if treatment doesn't work.



Caring for yourself and other family members

Your child's diagnosis will have an emotional effect on everyone in your family, as well as many friends and even acquaintances in the wider community. This section addresses your needs because, as a parent, your wellbeing is crucial to your child and the rest of your family.

Looking after yourself

Having a child diagnosed with kidney cancer has a huge impact on a parent or carer. You will likely have different feelings at different times, with periods of frustration, anger, fear, anguish, panic and grief. At some other times, you may feel quite calm, as you and your child settle into the routine of treatment.

You will probably find that your emotions go up and down a lot during the days and weeks following diagnosis, and that your feelings change over time. When you notice a difficult

or uncomfortable emotion, try to calm yourself and notice what you are feeling, rather than pushing it away. Sometimes, your feelings might spur you to take action or make a change. Other times, talking things through with someone you can trust is all that is needed.

One of the best things you can do for your family is to take care of your own needs. You can do this by eating and sleeping well, getting fresh air outside, addressing any health problems and taking regular breaks. By meeting your own needs, you can be there to help everyone else.



Supporting your child

The effect of a cancer diagnosis and treatment will depend on the age of your child. As well as feeling unwell and coping with side effects, they may be missing home, family and friends, and may struggle with the change in routine. If they are not able to say how they are feeling, they may express this through behaviour such as clinginess, tantrums or tearfulness.

One of the hardest parts of caring for a child with cancer is knowing what to say and how much information to give them. Many families feel that answering questions honestly is best, giving a little information at a time. Some children may not ask questions, but this doesn't mean they don't want to know what's happening. They may be frightened and uncertain of many things. Some children may even wonder if they have done something wrong and that's why they have cancer.

You can ask your child's medical team for guidance on how to talk to your child. There are also booklets for young children available from CCLG and Young Lives vs Cancer (YLVC) that can help you explain the illness and treatment. Younger children may be frightened about being separated from their parents. It's important to reassure them that any separation is only temporary. Doctors and nurses will be happy to explain more about this and can help you reassure your child.

Supporting siblings

Brothers and sisters of a child with cancer may have many or all the same feelings and emotions that you have. If you need to spend a lot of time in the hospital with your child, your other children may have to be cared for by family members or friends. They may have to spend a lot of time away from you and find their daily routine keeps changing. As well as worrying about their sibling's health, they may also feel resentful of all the attention they're getting. This can make them feel left out and angry. Being there for them or showing that you still love them can help to reassure and comfort them.

CCLG has a range of information to help brothers and sisters of children with cancer, from a storybook and animation for young children, to a parents and carer resource and a guide for older children and teenagers.



The CCLG booklet 'My brother or sister has 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

Animation for children:

Our short animation, based on the booklet is available to watch online at cclg.org.uk/my-brother-or-sister-has-cancer



Supporting your child at school

As your child's health improves, they may be able to go back to school. This is important for their educational, psychological, and social development, but it is also important for the whole family as school routines can help everyone return to a sense of 'normal life'. As soon as your child is diagnosed, contact your nursery or school's head teacher to tell them what's happening and keep open the lines of communication. It can help to let them know about the plans for treatment.

Many children diagnosed with kidney cancer are very young and have not yet started school. As a parent, you may have to choose between having your child at nursery throughout treatment or keeping your child at home. Keeping your child at home may mean they have less chance for social growth and development, but if they stay at nursery you may feel there is a risk of infection.

There is no right or wrong decision – it's a personal choice for you to make. You may want to think about whether your child:

- is already settled at nursery or pre-school
- has their social needs met by siblings and/or other children outside of the nursery
- is well enough to attend nursery or pre-school
- has already had chickenpox which can be harmful for children on treatment

It may be useful for you to talk to your specialist nurse or social worker about nursery attendance and the support they can offer to help with this.



The CCLG booklet 'Supporting your pupil after a cancer diagnosis' is an information guide for teachers but it is useful for parents and carers too.

Supporting grandparents

Being told that their grandchild has cancer will be a huge shock for your parents. They will worry not only about their grandchild but also how you will deal and cope with this news. Most are also concerned about the effects it will have on any other children within your family and, of course, how they will cope themselves.

As parents, you will usually have access to doctors and others who can answer your questions. It is not so easy for your parents to get information first hand and this can lead to feelings of stress and isolation. Keeping them involved and allowing them to help you and your family if they are able to can help them play a valuable role in supporting their family.

The CCLG booklet 'Supporting your grandchild and family' 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



Seeking information

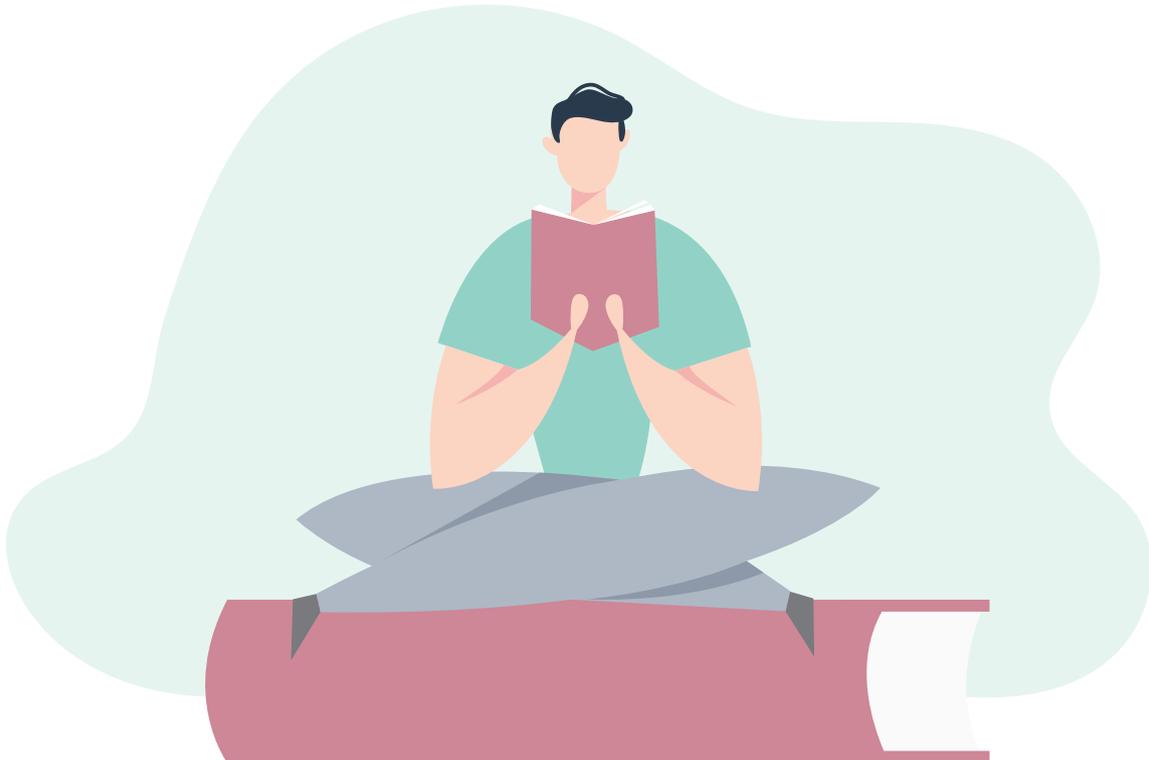
You may want to find out as much as possible about the cancer and its treatment. There is a lot of online information but not all of it is reliable, so talk to the doctors about where to look.

The CCLG leaflet 'Searching for information and support online' offers advice and tips on searching for cancer information on the internet.
www.cclg.org.uk/publications



The CCLG childhood cancer information hub brings together reliable information and resources about childhood cancer from a range of organisations into one, easy-to-search place www.cclg.org.uk/infohub

For more detailed information on caring for yourself and others, please read our accompanying booklet 'A parent's guide to cancer'.





Tumour cell types and glossary

Definitions of Wilms tumour cell types, subtypes, and other kidney tumours of childhood

Wilms tumours (also known as nephroblastoma) are made up of different types of cells. The classic histological (cell-type) pattern in Wilms tumour is made up of three key elements: epithelial, blastemal, and stromal. Around 5% of Wilms tumours are multifocal (containing one or more sub-type) and these tumours can be more difficult to assess. Kidney tumours are often categorised into low-risk, intermediate-risk and high-risk dependant on their histological make up:

Low-risk	Intermediate-risk	High-risk
Mesoblastic nephroma	Nephroblastoma - epithelial type	Nephroblastoma - blastemal type
Cystic partially differentiated nephroblastoma	Nephroblastoma - stromal type	Nephroblastoma - diffuse anaplasia
Completely necrotic nephroblastoma	Nephroblastoma - mixed type	Clear cell sarcoma of the kidney
	Nephroblastoma - focal anaplasia	Rhabdoid tumour of the kidney
	Non-anaplastic nephroblastoma and its variants	

Low-risk tumours

Mesoblastic nephroma

There are three cell (histological) sub-types of mesoblastic nephroma: the classical, cellular and mixed type. The difference between the three types has no effect on therapy.

Cystic partially differentiated nephroblastoma (CPDN)

CPDN is a distinct variant of nephroblastoma that usually occurs in children less than two years of age and treated with surgery only.

Completely necrotic nephroblastoma

Pre-operative chemotherapy may mean that the tumour is 'necrotic' or 'dead' when analysed under the microscope. Completely necrotic nephroblastoma following chemotherapy has been shown to have an excellent prognosis.

Intermediate-risk tumours

In order for tumours to be classified as intermediate-risk, the tumour has to be at least two-thirds (66%) of a particular sub-type under the microscope.

Nephroblastoma – Epithelial type

Epithelial cells are a type of cell that lines the surfaces of your body, e.g. skin, blood vessels. Epithelial nephroblastoma usually occurs in

younger children (average age 15 months) and are mostly classified as stage 1.

Nephroblastoma – Stromal type

Stromal cells are connective tissue cells found in any organ of the body. They help to support the function of the organ, i.e. kidney. Stromal cells often remain after pre-operative chemotherapy and stromal-type nephroblastoma usually occurs in younger children.

Nephroblastoma – Mixed type

In nephroblastoma of mixed type, there is a combination of blastemal and/or epithelial and/or stromal cells but none of them make up more than two-thirds of tumour. There can also be tumours which have more than 10% of blastema, regardless of the amount of epithelial or stromal components.

Nephroblastoma – Regressive type

This type is a tumour in which chemotherapy-induced changes have affected more than two-thirds of the tumour. The appearance of this type of tumour under the microscope is often a mixture of necrosis, fibrous stromal cells and sometimes evidence of a haemorrhage (blood loss).

Nephroblastoma – Focal anaplasia

This type is nephroblastoma which contains one or two 'foci' of anaplasia (anaplasia that can only be seen under a microscope). If the underlying cell make up of the tumour is blastemal, it should be classified as a high-risk tumour.

High-risk tumours

Nephroblastoma - Blastemal type

Nephroblastomas where at least two-thirds of the tumour consists of blastema are high-risk. Blastemal cells are called either undifferentiated, round or elongated cells, which are usually closely packed together under the microscope. Blastemal cells have different patterns under the microscope which include diffuse, serpentine, nodular, and basaloid patterns. The presence of blastema after pre-operative chemotherapy suggests non-response to treatment.

Nephroblastoma with anaplasia

Anaplasia may occur in the blastemal, epithelial or stromal components of nephroblastoma and it can be focal or diffuse. Focal anaplasia is defined as the presence of one or two clearly marked areas when looking under the microscope. Diffuse anaplasia is classed as anaplasia beyond the original tumour capsule or in the surrounding vessels and/or structures of the kidney.

Anaplasia occurs in about 5-8% of patients with nephroblastoma. Anaplasia never occurs in the first six months of life and it is very rare between 6-12 months (1-2%).

Nephrogenic rests

Nephrogenic rests are groups of embryonic kidney cells which persist after 36 weeks of pregnancy. They are usually only visible under the microscope in the kidney tissue next to the Wilms tumour, but sometimes they may form one or more small lumps visible on scans. They are considered to be potential pre-cancer cells for nephroblastoma. They have been found in 35-40% of patients with nephroblastoma.

Two main types of nephrogenic rests have been recognised: perilobar (A) and intralobar (B) rests. The rests may either regress to become fibrous tissue or progress to become nephroblastoma. Perilobar nephrogenic rests are linked with hemihypertrophy and Beckwith-Wiedemann syndrome, whereas intralobar rests are associated with WAGR and Denys-Drash syndromes. Nephroblastomatosis is the term used to describe the presence of persistent and multiple nephrogenic rests.

Glossary

Alopecia

Loss of hair.

Anaesthetic

Drug which stops feeling, especially pain. A general anaesthetic makes you unconscious. A local anaesthetic stops feeling in one part of the body.

Anaplastic Wilms tumour

About 5-10% of Wilms tumours have an appearance called anaplasia, which means the cells look very disorganised under a microscope. Considered to be a 'higher risk' type of Wilms tumour.

Bilateral

Affecting both kidneys.

Biopsy

Removal of a small piece of tissue for examination, to establish a diagnosis.

Blastemal-type Wilms tumour

This group of high-risk tumours can only be identified when chemotherapy has been given before surgery and by looking at the whole of the removed tumour. When a particular type of early kidney cell (blastemal cells) survive the pre-surgery chemotherapy in large numbers, the Wilms tumour is classified as 'blastemal-type'.

Bone marrow

The substance at the centre of long bones that makes blood cells.

Catheter

Tube passed into the body to drain fluid.

Central line

(Hickman line, Port-a-cath) Long plastic tube that is inserted into a large vein near the heart under anaesthetic. Central lines are used to take blood samples and give drugs.

Chemotherapy

Treatment using one or more anti-cancer drugs.

CT scan

Computerised Tomography scan. Multiple x-rays are taken by a CT scanner and converted by a computer to form a 3D view of the part of the body under examination.

Genetic

A condition caused by abnormal genes (may be inherited).

Haematuria

Blood in the urine.

Histology

The appearance of tissue under a microscope, which helps to determine the diagnosis.

Immune system

The body's defence against infection, disease and foreign substances.

Immunology

The study of the body's immune system, which fights infection.

Immunosuppressive

Lowering the body's ability to fight infection.

Intravenous (IV)

Into a vein, for example, when drugs are given directly through a drip.

Malformation syndromes

Very rarely, children 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).

Malignant

Cancerous.

MDT

Multi-disciplinary team. A group of doctors and other health professionals with expertise in childhood cancer, who together discuss the best course of treatment for their patients.

Metastases

Cancer that has spread from the place where it started (also known as secondary cancer).

MRI scan

Magnetic Resonance Imaging scan. An MRI scan relies on magnetism and is a very safe procedure as no radiation is used. It takes longer than a CT scan and is quite noisy.

Nausea

Feeling sick.

Nephrectomy

Surgery to completely remove a kidney. When less than the entire kidney is removed, this is known as partial nephrectomy or nephron-sparing surgery.

Nephroblastoma

Another name for Wilms tumour.

Neutropenia or neutropenic

Low levels of neutrophils, a type of white blood cell which fights infection.

Oncologist

A doctor who specialises in the treatment of cancer.

Oncology

The study and treatment of cancer.

Paediatric

To do with children.

Palliative

Relief of a symptom (for example, pain) rather than cure of the disease.

Prognosis

The outlook or expected outcome of a disease and its treatment.

Radiotherapy

The use of radiation to treat cancer.

Refractory

Resistant to treatment.

Relapse

The return of symptoms of a disease after a period of good health; recurrence of a tumour after treatment.

Remission

A period of good health where there is no longer any visible cancer.

Surgery

An operation.

Stem cell

Early (immature) blood cell from which other blood cells are made.

Therapy

Treatment.

Tumour

An abnormal lump of tissue formed by a collection of cells. It may be benign (non-cancerous) or malignant (cancerous).

Ultrasound scans

The sound waves produced by a scanner bounce from solid organs inside the body and are recorded on a screen. Allows doctors to see the outlines or shadows of normal organs and tumours.

Unilateral

Affecting one kidney only.

Help and support

Children's Cancer and Leukaemia Group (CCLG)

www.cclg.org.uk

Tel: 0333 050 7654

Information on childhood cancer, funding of research, and a full range of award-winning patient information resources, including Contact magazine.

Join our closed Facebook group just for parents and carers of a child with cancer:

www.facebook.com/groups/cclgparentcarergroup



Cancer Research UK

www.cancerresearchuk.org

Information on all cancer types and a key funder of research into cancer.

Young Lives vs Cancer

www.younglivesvscancer.org.uk

FREE helpline: 0300 330 0803

Advice and support for families affected by childhood cancer.

Macmillan Cancer Support

www.macmillan.org.uk

FREE helpline: 0808 808 0000

Practical, financial and emotional support for anyone affected by cancer.

The Little Princess Trust

www.littleprincesses.org.uk

FREE helpline: 0808 808 0000

Provides free real hair wigs to children with hair loss and funds research into childhood cancers.

Notes:

A series of horizontal dotted lines for taking notes.



the EXPERTS
in CHILDHOOD
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.

CCLG publications on a variety of topics related to children's cancer are available to order or download free of charge from our website. If you have any comments on this booklet, please contact us at publications@cclg.org.uk.

Our work is funded by donations. If you would like to help, text '**CCLG**' to **70300** to donate **£3**. This costs £3 plus a standard rate message.

Children's Cancer and Leukaemia Group
Century House
24 De Montfort Street
Leicester LE1 7GB

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

0333 050 7654
info@cclg.org.uk
www.cclg.org.uk

 ChildrensCLG
 CCLG_UK
 CCLG_UK

