Grace Kelly Childhood Cancer Trust

Renal rhabdoid tumours

Information for parents and carers

This publication is intended to supplement the advice given by your child's medical team. It was written by Dr Jennifer Kelly, General Practitioner and CEO of the Grace Kelly Childhood Cancer Trust.

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If you would like information on the references used to make this publication, please contact us.

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About this booklet

If you are reading this booklet, the likelihood is that your child, or a child close to you, has recently been diagnosed with a renal rhabdoid tumour. This booklet is designed to be a summary of some of the information you may have already been given and it may answer some of your questions. If you have any further worries or queries, please discuss them with the healthcare team looking after your child.

What is a rhabdoid tumour?

A rhabdoid tumour is a rare childhood cancer which may start in the kidneys, brain, spine or other parts of the body. There are 3 main types of rhabdoid tumours grouped together by the locations in which they originate:

1. **Renal malignant rhabdoid tumours (MRT)** - these occur or originate in the kidney (renal).

2. **Extra renal rhabdoid tumours (ERRT)** - these occur elsewhere in the body, such as in the liver, lungs or skin.

3. **Atypical teratoid rhabdoid tumours (AT/RT)** - these affect the brain and spinal cord (central nervous system).

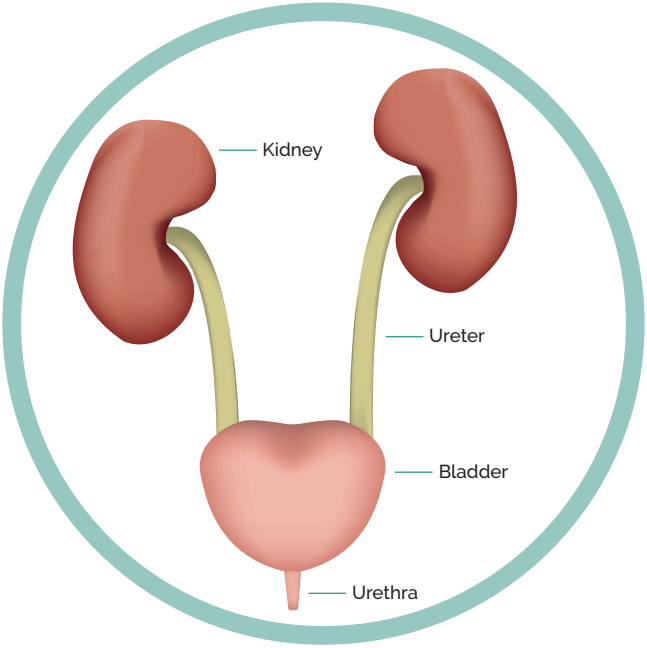
Renal rhabdoid tumours

Renal rhabdoid tumours are tumours that occur in the kidney (renal). They are very rare tumours that mostly affect infants (children less than 12 months of age) and very young children. The average age of diagnosis is 15 months old, however, they can occasionally occur in older children too.

What do the kidneys do?

There are two kidneys, one on each side of the spine just below the ribcage. The kidneys clean the blood by taking out waste and fluids as urine.

The urine produced then passes through the ureter into the bladder and out of the body. The kidneys also make hormones that help control blood pressure and signal the bone marrow to make red blood cells when needed.



Symptoms of renal rhabdoid tumours

Children with renal rhabdoid tumours may have experienced some of the following (but others may have very few or no symptoms):

- A swollen tummy, a visible lump or a fullness on the side of the tumour.
- Very young children may be fussier or clingier than normal.
- Older children may complain of pain.
- Visible blood in their urine. This could be fresh blood (looks pink or red) or old blood (makes the urine a darker brown cola colour).
- Recurrent high temperatures.

When examined by a nurse or doctor, children may also have high blood pressure.

What makes it a renal rhabdoid tumour?

Often, on scans, a renal rhabdoid tumour looks similar to other renal (kidney) tumours such as Wilms Tumours. It is not until the cells of the tumour are examined under a microscope that a renal rhabdoid tumour diagnosis can be made.

Almost all renal rhabdoid tumours have a characteristic genetic change in the cells called a SMARCB1 (or INI1) mutation. It is this change that is responsible for the development of malignant rhabdoid tumours.

For more information, please see our information booklet ***"The genetics of rhabdoid tumours."***

Staging of renal rhabdoid tumours

Staging is an assessment that is made by doctors for all patients with cancer to help them plan their treatment. It categorises whether the tumour is in a single place or whether it has spread elsewhere in the body.

Renal rhabdoid tumours are classified as:

Stage I: The tumour can only be seen in the kidney and there are no signs of spread. The tumour wall (the capsule) is not damaged, and the tumour has been removed whole.

Stage II: The tumour may have extended beyond the kidney but has been removed. Some tumour cells may be left behind but there is no spread to the lymph nodes.

Stage III: The main tumour may have spread to local lymph nodes or other local parts of the body or some of the tumour is left behind after surgery.

Stage IV: The tumour has spread to distant lymph nodes or organs. Often this may be in the lungs, liver and brain. This is the most common stage for children to have when they are diagnosed.

Stage V: Tumours are present in both kidneys.

What investigations are needed?

The following investigations may be required:

- Ultrasound scan.
- **Computerised tomography** (CT) scan.
- **Magnetic resonance imaging** (MRI) scan - usually including the abdomen (tummy), chest and head.
- Biopsy of the tumour.
- Blood tests.
- Chest X-ray.
- **ECG** (heart tracing).
- **Echocardiogram** (heart scan).

A number of children with renal rhabdoid tumours have signs that their tumour has spread to other parts of the body at diagnosis. This may make it difficult to remove all of the tumour.

Treatment of children with renal rhabdoid tumours

If your child has a renal rhabdoid tumour, the child will receive their treatment in a specialist care centre that is experienced in treating children with cancer. Most children will be offered a combination of surgery, chemotherapy and often radiotherapy depending on the tumour location and the age of the child.

Although renal rhabdoid tumours are rare, there are standard treatment guidelines in the UK.

If your child has relapsed or has a tumour that has not responded fully to treatment, you may be offered the opportunity to take part in a clinical trial of a new medication. Your doctor and members of the care team will discuss the options with you in depth.

Surgery

If your child is less than 6 months old, the initial treatment may be to remove the whole kidney with the tumour inside it.

If your child is over 6 months of age, one of two things may happen:

i) A biopsy (taking a small piece of the tumour) is often done first. This is then followed by chemotherapy and then surgery to remove the whole kidney.

ii) Sometimes, chemotherapy may be given before a biopsy is taken because, on scans, rhabdoid tumours can appear similar to other types of tumours (including Wilms tumours) which are treated in a slightly different way (chemotherapy (without biopsy) followed by surgery to remove the whole kidney and tumour together).

Removing the whole tumour is always the aim of surgery, but this may not be possible straight away.

If your child's tumour is in a difficult location or is too large to remove surgically, chemotherapy is often given first to help shrink the tumour.

Chemotherapy

This is a cancer treatment in which medications are used to kill cancer cells and shrink tumours. Rhabdoid tumours can be challenging to treat because they have a tendency to become resistant to chemotherapy (the chemotherapy stops working as well).

To help reduce the chances of this happening, a combination of chemotherapy drugs are given, often in alternating cycles. This helps to fight the tumour in the most effective possible way.

Radiotherapy

Children who develop rhabdoid tumours may also receive radiotherapy as part of their treatment, however, it may not be suitable in all situations.

Supportive care

This is an important part of treatment for children with rhabdoid tumours. Supportive care means helping to keep the child as comfortable and as free from symptoms as possible during their treatment. It includes treatment for infections, pain relief and medication to reduce side effects of the treatment they are receiving such as medications for sickness. Children may also require some nutritional support (help with feeding).

In many areas as a matter of routine, all children with cancer are referred to the local children's hospice team to help with supportive care (symptom management) whilst they are receiving active treatment.

After treatment: What happens next?

On completing treatment, the frequency of your child's appointments will decrease, but they will continue to receive regular follow up. This will usually consist of an examination, blood tests, an MRI scan to detect any recurrence and for some children, other investigations too.

After treatment, children may face a range of challenges including local effects resulting from the tumour itself as well as the effects of the treatments that they have undergone.

Is my child's treatment going to work?

Statistics can tell us the average outcomes of all children diagnosed with renal rhabdoid tumours. They tell us the proportion of children we would expect to do well, and the proportion of children expected to not do so well. However, it is impossible to predict how each individual child will do.

What we do know:

- Renal rhabdoid tumours are challenging to treat and do not have good survival rates on average.
- Children with no signs of tumour spread at diagnosis tend to do better on average than those with tumour spread.
- Children who are over 24 months of age at diagnosis are likely to do better on average.
- We cannot predict the exact outcome of each child.

This is why it is important to remember that your child is unique and may not follow the expected course of treatment or outcome. Every child is different.

Your child's consultant and medical team will help give you advice and information to make any decisions needed. If you have any concerns or questions, please speak to a member of your child's team.

Advice for parents and carers

If your child has recently been diagnosed with a renal rhabdoid tumour, you may well be feeling overwhelmed. Many parents talk about going into survival mode, but it is important to take time to look after yourself when you can.

Talking to loved ones or professionals and asking for practical help when needed can make a difference. If you do not know where to turn, please speak to your child's medical care team or your General Practitioner for more support and advice.

For more information on booklets we produce and for a link to our online rhabdoid parent's support group, please see **www.gkcct.org**

Grace Kelly Childhood Cancer Trust

The Grace Kelly Childhood Cancer Trust is a UK-based children's cancer charity that concentrates on funding research and providing support to families. We teach the signs and symptoms of childhood cancer and are working towards earlier diagnosis of children through the education of parents, carers and clinicians.

Our medical publications are written by medics and are extensively peer-reviewed. Bridging the gap between parents and medics.

For more information on our work and the publications we have produced, please see our website.

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