Grace Kelly Childhood Cancer Trust

Extrarenal 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.

Our thanks go to Professor Bernadette Brennan (Professor of Paediatric Oncology, Manchester), Dr Stephen Lowis (Consultant Paediatric Oncologist, Bristol), Professor Kathy Pritchard-Jones (Professor of Paediatric Oncology, UCL), Dr Mette Jorgensen and Dr Olga Slater (Consultant Paediatric Oncologists, GOSH), Dr Patricia O'Hare (Consultant Paediatric Oncologist, Royal Belfast Hospital) and Dr Sam Behjati (Group Leader at the Wellcome Sanger Institute and Consultant Paediatric Oncologist, Addenbrooke's) for their help in the peer review of this resource.

The Grace Kelly Childhood Cancer Trust has made every effort to ensure information is accurate at the time of printing. **We are proud to be a certified member of The PIF TICK (Trusted Information Creator), the only UK-wide quality accreditation mark for print and online health information.**

This means you can be assured that what you are reading, watching, or listening to is evidence-based, understandable, jargon-free, up-to-date, and produced to the best possible standard.

We are unable to accept responsibility for any information provided by third parties.

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

The publication of this booklet was funded by the Grace Kelly Childhood Cancer Trust.

Published June 2026, review due June 2029.

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 an extrarenal rhabdoid tumour (ERRT). 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 that can 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 first develop:

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

2. **Extrarenal rhabdoid tumours (ERRT)** - these occur elsewhere in the body, such as in the liver, lungs, skin and other soft tissues.

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

About extrarenal rhabdoid tumours

Extrarenal rhabdoid tumours (ERRT) are very rare tumours that can occur in any part of the body other than the kidney, for example, the liver, lungs, skin and muscle.

They are usually found in infants and young children but can occur in older children and occasionally in adults.

Symptoms of extrarenal rhabdoid tumours

Extrarenal rhabdoid tumours can present with a lump anywhere in the body and therefore can cause a variety of symptoms. Sometimes, if located in soft tissue or bone, it may cause a child to have a lump or swelling that is increasing in size.

Children with ERRT may also develop one or a few of the following symptoms:

- Feeling tired.
- A swollen tummy.
- Vomiting.
- Paralysis or weakness of the muscles of an arm, leg or the face.
- Unsteady walking or loss of balance.
- Problems with breathing, for example, fast or laboured breathing.
- Pain in the area the tumour is located.

What makes it an extrarenal rhabdoid tumour?

Often, on scans, extrarenal rhabdoid tumours can look similar to other types of tumours. It is not until the cells of the tumour are examined under a microscope that a diagnosis of ERRT can be made.

ERRTs have a characteristic genetic change in the cells called a SMARCB1 (or INI1) mutation. This change is identified when cells of the tumour are examined under a microscope. 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 extrarenal rhabdoid tumours

Staging simply means an assessment that is made by doctors for all patients with cancer. This helps them to plan treatment. Staging categorises the tumour by its location and whether there are any signs that the tumour has spread.

Stage I: The tumour can only be seen in one place (its origin) and there are no signs of spread. The outside wall of the tumour (the capsule) is not damaged, and the tumour has been removed whole without being biopsied.

Stage II: The tumour can only be seen in one place and there is no spread to the lymph nodes. Some of the tumour has extended further than the tissues it started in, meaning some cells may be left behind when it is removed.

Stage III: The main tumour may have spread to local lymph nodes or other local parts of the body. The tumour has been biopsied or most of the tumour is left behind.

Stage IV: The tumour has spread to distant lymph nodes or organs. Often this may be in the lungs, liver and brain.

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).

Some children with ERRT have signs that their tumour has spread to other parts of the body. This may make it difficult to remove all of the tumour.

Treatment of children with extrarenal rhabdoid tumours

If your child has an extrarenal rhabdoid tumour, their treatment will take place 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 ERRTs 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, they may be offered the opportunity to take part in a clinical trial of a new medication. Your doctors and members of your child's care team will discuss the options with you in depth.

Surgery

After biopsy, surgery is usually the first step of treatment. Depending on the size and location, removal of the whole tumour may not be possible. If the child's tumour is in a difficult location or is too large to remove surgically, chemotherapy may be 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. Extrarenal rhabdoid tumours can be challenging to treat because they have a tendency to become resistant to chemotherapy (the chemotherapy stops working as well).

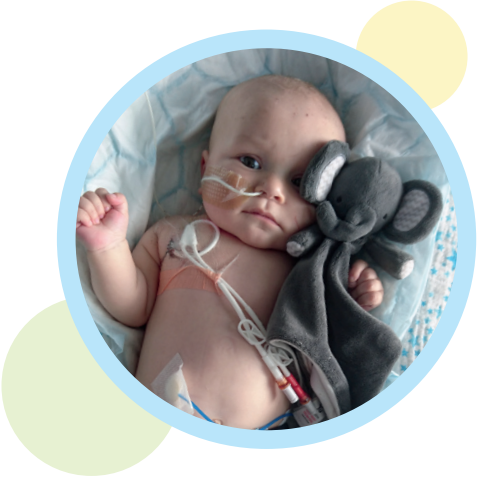
To help reduce the chances of this happening, a combination of different chemotherapy drugs are given, often in alternating cycles. This helps to fight the tumour in the most effective way possible.

Radiotherapy

Children over 6 months of age who develop extra renal rhabdoid tumours may also receive radiotherapy as part of their treatment, but this depends on the position of the tumour and a clinical assessment of your child.

Supportive care

This is an important part of treatment for ERRT. Its role is to help keep the child as comfortable and as free of 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.



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 on 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 usually consists 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 caused either by the tumour itself or as a result of the treatments that they needed to receive. Follow-up appointments can be useful for addressing these challenges should they occur.

Is my child's treatment going to work?

Statistics can tell us the average outcomes of all children diagnosed with extrarenal rhabdoid tumours. They will 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 know:

- Extrarenal 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 signs of tumour spread.
- Children who are over 12 months of age at diagnosis tend to do better on average than those under 12 months at diagnosis.
- 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 an extrarenal rhabdoid tumour, you may well be feeling overwhelmed with the whole situation. 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 by asking for practical help when you need, 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.

Grace Kelly Childhood Cancer Trust

www.gkcct.org

gracekellychildhoodcancertrust

gracekellychildhoodcancertrust

@GraceKellyTrust

Registered charity number 1167783.

Trusted Information Creator

Patient Information Forum