Extra Renal Rhabdoid tumours

Information for parents and carers

Grace Kelly Ladybird Trust

This publication is intended to supplement the advice given by your medical team. It was written by Dr Jennifer Kelly, GP and founder of the Grace Kelly Ladybird Trust.

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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 an extra renal rhabdoid tumour. This leaflet is designed to be a summary of some of the information you may have been already 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 part of the body. There are 3 main types of rhabdoid tumours, grouped together by the locations in which they originate:

1. Malignant renal 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, skin and other soft tissues.
3. Atypical teratoid / rhabdoid tumours (AT/RT) - these affect the brain and spinal cord (central nervous system).

Confusingly, the term malignant rhabdoid tumour is sometimes used as a general term to describe extra renal rhabdoid tumours as well. For example, some consultants may refer to a liver (hepatic) rhabdoid tumour as a malignant rhabdoid tumour of the liver or as a hepatic extra renal rhabdoid tumour.

About extra renal rhabdoid tumours

Extra renal rhabdoid tumours are rare tumours that can occur in any part of the body other than the kidney, for example the liver, lungs, skin and muscle.

ERRTs are usually found in infants and young children, but can occur in older children and occasionally in adults. There are around 3 cases of extra renal rhabdoid tumours diagnosed each year in the UK.

Symptoms at diagnosis

Extra renal rhabdoid tumours can present with a lump anywhere in the body and hence can cause a variety of symptoms.

Sometimes an extra renal rhabdoid tumour in soft tissue or bone may present with a **lump or swelling** that is increasing in size.

It may also cause some of the following depending on the location of the tumour:

- Feeling tired
- Pain
- Vomiting
- Paralysis or weakness of the muscles of an arm, leg or the face
- Unsteady walking or loss of balance
- Abdominal (tummy) swelling
- Problems with breathing, for example fast or laboured breathing

What makes it an extra renal rhabdoid tumour?

Extra renal rhabdoid tumours 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 (or mutation) 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 extra renal rhabdoid tumours

Staging is an assessment made by doctors for all patients with cancer to help plan treatment. It categorises the tumour by its location and whether it has spread to any nearby or distant locations in the body.

Extra renal rhabdoid tumours grow quickly and spread (metastasise) early, therefore many children already have signs of tumour spread at diagnosis.

Stage I: The tumour is limited to its site of origin. The tumour wall (the capsule) has not been damaged and it has been removed whole. The tumour has not been biopsied prior to it being removed.

Stage II: The tumour may have extended beyond the original tissues of origin and has been removed leaving some tumour cells behind. There is no spread to the lymph nodes.

Stage III: The main tumour may have spread to local lymph nodes and/or other local structures. 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**
- **CT** (computerised tomography) and/or
- **MRI** (magnetic resonance imaging) scan – usually including the abdomen (tummy), chest and head.
- **Biopsy** of the tumour
- **Blood tests** including kidney tests (GFR)
- **Bone scan**
- **Chest X-ray**
- **ECG** (heart tracing)
- **Echocardiogram** (heart scan)

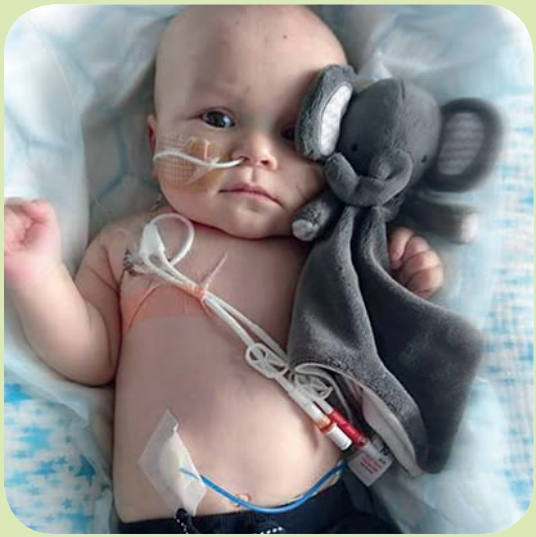
Treatment of extra renal rhabdoid tumours

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 extra renal rhabdoid tumours are rare, there are standard treatment guidelines in the UK. If your child has a relapsed tumour, or one that has not responded fully to treatment, you may be offered the opportunity to take part in a clinical trial of a new drug. Your doctor and members of the 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. Extra renal rhabdoid tumours are typically aggressive and can become resistant to chemotherapy quickly. To help reduce this possibility, a combination of chemotherapy drugs are given, often in alternating cycles to help fight the tumour in the most effective possible way.

Radiotherapy

Children over 6 months of age who develop extra renal rhabdoid tumours may also receive radiotherapy as part of their treatment.

Supportive care

This is an important part of treatment for extra renal rhabdoid tumours. Its role is to help keep the child as comfortable and as free of symptoms as possible. It includes treatment for infections, pain relief and treatments to reduce side effects such as 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 - follow up

On completing treatment, the frequency of appointments will decrease but there will continue to be regular follow up. This will usually consist of an examination, blood tests and an MRI scan to detect any recurrence.

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. The Children's Cancer and Leukaemia Group offer a range of booklets that are very helpful for further information.

Extra renal rhabdoid tumours - long term

Statistics tell us the average outcome for children with the same condition. They indicate the proportion of children who would be expected to do well, and the proportion who would be expected not to do so well. However, it is impossible to predict how each individual child will do. This is why it is important to remember your child is unique and may not follow the expected course of treatment or outcome.

What we know:

- Unfortunately, overall, extra renal rhabdoid tumours do not have good survival rates.
- Outcomes are poorer if a child has signs of tumour spread at diagnosis.
- Children under 12 months of age who develop extra renal rhabdoid tumours are less likely to do as well as older children.
- We cannot predict the exact outcome of each child.

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.

Grace Kelly Ladybird Trust

The Grace Kelly Ladybird Trust is a UK children's cancer charity that concentrates on funding research and support for children with rare solid tumours. We also work to provide education on the signs and symptoms of childhood cancer and how it may present.

For more information on rhabdoid tumours and a link to our online support group (available for parents of children affected by rhabdoid tumours) please see our website.

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