Grace Kelly Childhood Cancer Trust

Atypical teratoid 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 an atypical teratoid rhabdoid tumour (AT/RT). This booklet is designed to be a summary of the information you may have already been given and it may answer some of your questions. If you have any further 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 brain, spine, kidneys or other parts of the body. There are 3 main types of rhabdoid tumours, grouped together by the sites in the body in which they first develop:

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

2. **Renal rhabdoid tumours** - these occur in the kidney (renal).

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

Almost all rhabdoid tumours have a characteristic genetic change in the cells called a SMARCB1 (or INI 1) mutation. It is this change that is responsible for the development of rhabdoid tumours.

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

About atypical teratoid rhabdoid tumours

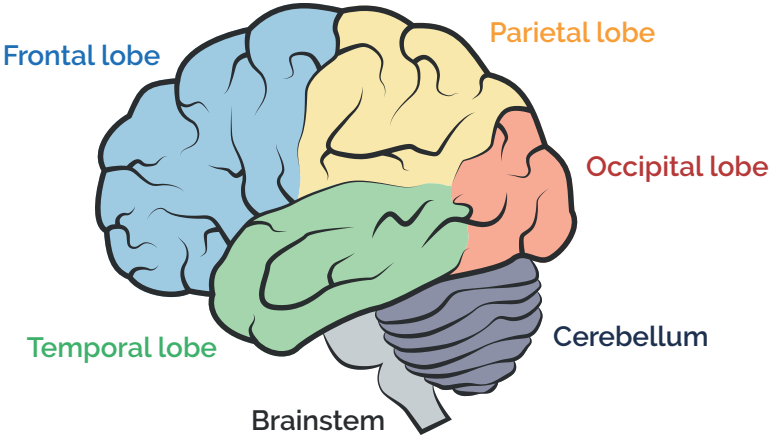
Atypical teratoid rhabdoid tumours (AT/RT) are very rare brain and spinal cord tumours, forming only about 1-2% of all brain tumour diagnoses in children. This means for every 100 brain tumours that are diagnosed in children of all ages, only one or two of them would be AT/RT.

AT/RTs can develop at any age; however, they occur most commonly in very young children and account for 1 in every 5 brain tumours that occur in children under the age of 3 years.

Atypical teratoid rhabdoid tumours often grow rapidly resulting in the development of symptoms over a relatively short period of time.

What makes it an atypical teratoid rhabdoid tumour?

Often, on scans, an atypical teratoid rhabdoid tumour looks similar to other brain and spinal tumours. It is not until the cells of the tumour are examined under a microscope that an AT/RT diagnosis can be made.



Symptoms of brain tumours

Children with brain tumours may have experienced some of the following:

- Headaches on waking in the morning, often accompanied by vomiting.
- Nausea and vomiting, especially in the morning.
- Being unusually sleepy or unable to do skills previously mastered.
- An increase in head size in babies or appearing unusually fussy.
- Blurred or double vision.
- In older children, problems with walking, coordination or losing balance.

If you notice your child developing new symptoms at any time, please do let your child's care team know. Their symptoms can vary depending on the exact location of their tumour in the brain, its size and a number of other factors.

Where in the brain do AT/RTs develop?

The majority of AT/RTs develop in the lower, back parts of the brain called the cerebellum and brain stem, but they can occur in other locations too.

Different locations of the brain have different functions which can be disturbed if a tumour develops in that area.

The cerebellum is important in the control of movement, posture and balance.

The brainstem controls the flow of messages between the brain and the rest of the body and is responsible for controlling basic bodily functions for example breathing, blood pressure and heart rate.

Staging of atypical teratoid rhabdoid tumours

The staging of a person's tumour is an assessment made by doctors to help plan their treatment. It is used to categorise whether the tumour is in a single place or whether it has spread elsewhere in the brain or the rest of the body.

There is currently no standard staging system for AT/RTs, but tumours are categorised either as:

- **Localised** - occurring in only one location in the brain.
- **Metastatic** - the tumour has spread to other parts of the brain or spinal cord.
- **Multifocal** - tumours develop in more than one location at the same time.

Certain investigations can help to determine whether all or part of the tumour can be removed during surgery.

What investigations are needed?

The following investigations may be required:

- **Computerised tomography** (CT scan).
- **Magnetic resonance imaging** (MRI) of the brain and spine.
- **Lumbar puncture** which removes a small sample of cerebrospinal fluid (CSF) from the spine using a special needle. This is then analysed to assess whether any tumour cells are present in the fluid removed.
- **Ultrasound of the abdomen** (tummy area) to check that other organs are healthy and there is no evidence of other tumours.
- **Blood tests.**
- **Echocardiogram** (heart scan).
- **Biopsy or resection** (removal) of the tumour to find out the type of tumour.

Sometimes other investigations may be required as well.

After the tumour (or parts of it) are removed or biopsied, it is examined and tested under a microscope to identify the tumour type.

Some children with AT/RT have signs that their tumour has spread to other parts of the brain or spinal cord at diagnosis. This may make it difficult to remove all of the tumour.

Treatment of children with atypical teratoid rhabdoid tumours

If your child has an atypical teratoid 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 sometimes radiotherapy depending on the tumour location and the age of the child.

Although AT/RTs are rare tumours, there are standard treatment guidelines in the UK. The ATRT01 trial is due to open in the UK soon, so your child may be offered their treatment as part of a clinical trial.

If your child has relapsed or has a tumour that has not responded fully to treatment, you may also 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

This is often the first step in treating AT/RTs. The surgeon will always try to remove the tumour completely if possible. If not possible initially, a second operation may be possible after one, two or three blocks of chemotherapy.

Chemotherapy

This is a cancer treatment in which medications are used to kill cancer cells and shrink tumours. Atypical teratoid 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, chemotherapy for AT/RT is given very intensively and more frequently than for most types of tumour in order to get the tumour under control quickly.

High-dose chemotherapy with a stem cell transplant may also be given.

Radiotherapy

Children with atypical teratoid rhabdoid tumours may have radiotherapy, dependent on their age. Generally, radiotherapy to the brain and spine is avoided in children under 12 months of age and used with caution in children under 3 years of age due to the problems it can cause for very young children.

For children that have radiotherapy, it is usually given quite early – within 6–8 weeks of surgery.

Supportive care

This is an important part of treatment for children with AT/RTs. Supportive care means helping 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 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 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 and care. 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 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 atypical teratoid 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:

- AT/RTs 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 12 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 AT/RT, 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 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 the 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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