The ROYAL MARSDEN
NHS Foundation Trust

Ependymoma

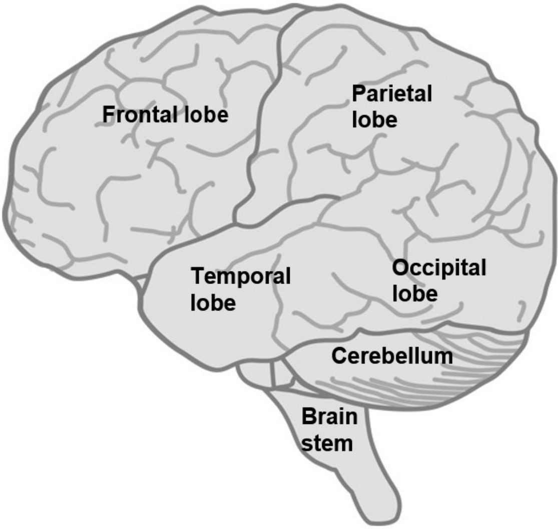
Children's Unit

Patient Information

NHS

What is an ependymoma?

An ependymoma is a type of tumour that develops from the cells which line the fluid-filled spaces of the brain and spine. They can be either malignant (cancerous) or benign (non-cancerous) depending on how quickly growing the tumour is. They can occur anywhere in the brain or spine and may spread to the spinal cord through the cerebrospinal fluid (CSF).



The brain

Who gets an ependymoma?

Ependymoma accounts for less than 10% of all childhood brain and spinal cord tumours and is more common in younger children. Ependymomas can occur at any age although it is rare in adults. Ependymomas are twice as likely to occur in the area at the lower back of the head (posterior fossa). Like most brain tumours, the cause of ependymoma is unknown.

What are the signs and symptoms?

The symptoms depend on where the tumour is and whether there is increased pressure in the head (raised intracranial pressure). Symptoms include:

- nausea and vomiting (most common)
- lethargy and irritability
- headaches
- clumsiness
- difficulty with tasks such as handwriting
- gradual decline in school work
- changes in personality and behaviour
- abnormal gait (the way the child walks).

If the tumour spreads or is located in the spinal cord, the signs and symptoms may include:

- back pain
- difficulty walking
- problems with bowel and bladder control.

What tests (investigations) will be carried out?

We will need to carry out some tests to find out as much as possible about the type, position and size of the tumour. This will help us to decide on the best treatment for your child. These tests include:

- **CT scan** – it is likely a CT scan of your child's brain was the first specific test carried out at your local hospital. Although MRI scans are usually the best way of seeing the tumour and the effects of treatment, sometimes CT scans are also useful. For more information, please see The Royal Marsden leaflet *CT scan*.

- **MRI scan** – this scan allows us to see the brain and spine in great detail and is used regularly to diagnose and follow the effect treatment is having on your child's tumour. For more information, please see The Royal Marsden leaflet *MRI scan*.
- **Lumbar puncture (CSF testing)** – this is carried out under a general anaesthetic about 14 days after surgery. We look for any malignant cells in the CSF.

Staging

Staging is a measure of how far the tumour has spread beyond its original site. The most important features in ependymoma are whether the tumour has spread to the spinal canal and if the neuro-surgeon has removed the entire tumour.

How do you treat an ependymoma?

The treatment used depends on location of the tumour and the age of the child. Common treatments include:

Surgery

All children will have an operation to confirm the diagnosis, relieve pressure in the head and to try and remove as much of the tumour as possible. Surgery is very important and in some cases where the neuro-surgeon cannot remove all of the tumour at the first operation, they will try and remove all the remaining tumour later.

Radiotherapy

Radiotherapy is often used after surgery to destroy any remaining cancer cells. If there is no evidence of spread through the CSF, radiotherapy is given locally to the site of the tumour. However, any evidence of spread will mean the whole brain and spine will need to be treated. In young children (especially under the age of three years), radiotherapy may be delayed, reduced or possibly avoided due to the significant effects of radiotherapy on a young child's brain. Please see The Royal Marsden booklet *Radiotherapy; your questions answered* for more details on this type of treatment.

Proton beam therapy

Children with ependymoma may benefit from receiving proton beam therapy. Proton beam therapy delivers radiotherapy in a way that may reduce the radiation to healthy tissue outside the area needing treatment. This can mean that children have fewer long-term side effects. In the UK, proton therapy is delivered at University College Hospital, London and The Christie Hospital, Manchester. If your child is eligible for NHS funding, you will be informed by the medical team and have the opportunity to discuss this in detail. Proton therapy is not available at The Royal Marsden.

Chemotherapy

Chemotherapy is usually given to shrink any remaining tumour so that further surgery may be carried out. It may also be given so that we can avoid or delay treatment with radiotherapy in young children. Treatment with anti-cancer drugs is used to destroy the tumour cells. It is usually given by injections and drips into a vein (intravenous infusion). The type of chemotherapy varies in intensity and duration depending on whether the tumour has spread, if radiotherapy will be used and the age of the child. If your child takes part in a clinical trial, the treatment is explained in more detail in the specific trial information sheet. We will give you an outline of the treatment in the form of a ‘road map’, known as a protocol.

What are clinical trials?

Clinical trials are medical research trials involving patients that are carried out to try and find new and better treatments. In cancer, clinical trials are most commonly used to try and improve different forms of treatment such as surgery, radiotherapy or chemotherapy.

The treatment being tested may be aimed at:

- improving the number of people cured (for example, trying new types of surgery or chemotherapy)
- improving survival

- relieving the symptoms of the cancer
- relieving the side effects of treatment
- improving the quality of life or sense of wellbeing for people with cancer.

Clinical trials may also involve research aimed at understanding more about the tumour's biology. You may be asked to allow us to do research on the tumour sample removed at surgery, or on blood samples.

Well-run clinical trials have led to a significant improvement in the treatment of children with cancer. You can find more general information in The Royal Marsden booklet *Clinical trials*. If you are asked to consider entering your child into a clinical trial, you will be given specific information about the trial before deciding whether to take part. There is no obligation to take part and the care your child receives will not be affected by your decision.

General side effects of chemotherapy

Bone marrow suppression (myelosuppression) – chemotherapy drugs decrease the production of blood cells by the bone marrow for a variable period of time. This results in low red blood cells (anaemia), low white blood cells (neutropenia) and low platelets (thrombocytopenia). Your child may need blood or platelet transfusions and will be at increased risk of infections. The doctors and nurses caring for your child will tell you more about these side effects.

Nausea and vomiting – some of the chemotherapy drugs used may make your child feel sick or vomit. We will give anti-sickness drugs at the same time to stop nausea and vomiting which are usually very effective.

Sore mouth (mucositis) – some of the chemotherapy drugs make the lining of the mouth and throat very sore and ulcerated. We will give your child painkillers for this, and explain how to care for your child's mouth during treatment.

Hair loss – temporary hair loss is common.

Weight loss – nausea, vomiting, mucositis and taste changes may result in your child losing weight. A dietitian will help you to support your child's nutrition during this time.

Chemotherapy drugs

For details of the side effects of individual drugs, please see the Macmillan individual drug information factsheets available from the Macmillan website or via your Clinical Nurse Specialist (CNS).

What is the outlook (prognosis)?

This is very closely related to whether the neuro-surgeon can remove all of the tumour, its spread and how quickly it grows (aggressiveness). Children who have not had all the tumour removed at initial surgery, or evidence of tumour spread, have a reduced survival rate.

What are the possible long-term effects?

As ependymomas can occur in young children and the treatment is given at an important time of the child's development, there may be some long-term effects of treatment. These could include growth and hormonal changes, reduced fertility, behavioural changes, possible learning problems and difficulties with coordination. Hearing and visual disturbances can result from both tumour and treatment. There is a small risk of developing a second cancer later in life.

All children are followed up after treatment is finished in the 'long-term brain tumour follow up clinic'. Your child will be seen at regular intervals in this clinic, indefinitely, so that we can help with any long-term effects of the treatment.

Contact details

Children and Young People's Unit
24 hour Helpline: **020 8915 6248**

Other sources of information and support

General information on radiotherapy and side effects is available in other booklets from the PALS Help Centre, via *www.royalmarsden.nhs.uk* and in the 'parent held record'.

Macmillan Cancer Support

Macmillan Support Line: **0808 808 0000**
Website: *www.macmillan.org.uk*

Macmillan provides a range of free information and support on all aspects of cancer including:

- advice on benefits and other kinds of financial support
- information on local cancer support groups and organisations near you.

The Children's Cancer and Leukaemia Group (CCLG)

Website: *www.cclg.org.uk*

CCLG is a national professional body responsible for the organisation of the treatment and management of children with cancer in the UK. They coordinate national and international clinical trials. They also provide information for patients and families and produce a quarterly magazine called Contact.

The Brain Tumour Charity

Website: *www.thebraintumourcharity.org*
Support and information line: **0808 800 0004**

The Brain Tumour Charity is a UK charity, committed to fighting brain tumours. They fund scientific and clinical research into brain tumours and offer support and information to those affected, whilst raising awareness and influencing policy.

Young Lives vs Cancer

Website: *www.younglivesvscancer.org.uk*

Support line: **0300 303 5220**

Young Lives vs Cancer is a UK charity for children, young people, and their families, which provides clinical, practical, financial and emotional support to help them cope with cancer.

Brains Trust

Website: *www.brainstrust.org.uk*

Brainstrust is a UK brain tumour charity which is dedicated to helping people affected by a brain tumour. They aim to provide support and advice from the point of diagnosis.

References

This booklet is evidence based wherever the appropriate evidence is available, and represents an accumulation of expert opinion and professional interpretation.

Details of the references used in writing this booklet are available on request from:

The Royal Marsden Help Centre

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Should you require information in an alternative format, please contact The Royal Marsden Help Centre.

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