The ROYAL MARSDEN
NHS Foundation Trust

Medulloblastoma / PNET

Children's Unit

Patient Information

What is a medulloblastoma/PNET?

A medulloblastoma / PNET is a malignant (cancerous) tumour formed from poorly developed brain cells. Most of these tumours originate in the lower back part of the brain (the cerebellum) and are called medulloblastomas. When they occur in other parts of the brain they are called PNETs (Primitive Neuro Ectodermal Tumours).

If a PNET occurs in the area of the brain called the pineal gland, then the PNET is known as a pineoblastoma. All PNETs can sometimes spread (disseminate or metastasise) to the spinal cord through the cerebrospinal fluid (CSF) so this will be important to assess – see investigations on page 3. They very rarely spread to other organs of the body.

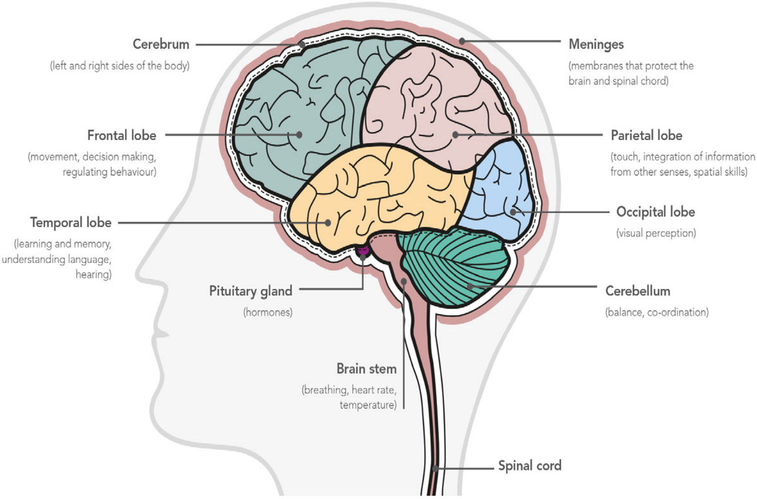

Image courtesy of Children's Cancer and Leukaemia Group

Who gets a medulloblastoma / PNET?

Public health records published in 2018 show that on average 349 children were diagnosed per year with a brain or spinal tumour. Of all the brain or spinal tumour diagnosis in the UK annually, approximately 15% (15 in 100 cases) are confirmed as medulloblastoma or PNET. This suggests approximately 70 new diagnosis of medulloblastoma per year. They are most common in children between the ages of three and eight, and are slightly more common in boys than girls. They can also be found in adults but are extremely rare. Like most brain tumours, the cause of medulloblastoma or PNET is usually unknown.

What are the signs and symptoms?

The symptoms are usually due to 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 to 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 that a CT scan of your child's brain was 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 used. 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** – this is carried out under general anaesthetic about 14 days after surgery, to look for any malignant (cancerous) cells in the CSF.

How do you treat a medulloblastoma / PNET?

The treatment used depends on where the tumour is, if it has spread and the age of your child. Common treatments include

Surgery

All children will have an operation to confirm the diagnosis, relieve pressure and try to remove as much of the tumour as possible.

Radiotherapy

Radiotherapy is often used after surgery to destroy any remaining malignant cells. As medulloblastoma / PNET may spread through the CSF to the spinal cord, radiotherapy is given to the brain and spinal cord. However, in young children (especially under the age of three years), we may delay, reduce or possibly avoid radiotherapy. This is because of the significant effects of radiotherapy on a younger child's brain. Please see the Children's Cancer and Leukaemia Group (CCLG) leaflet *Radiotherapy treatment* for more details on this type of treatment.

Radiotherapy may be given as proton therapy which is currently available on the NHS through hospitals in Essen, Germany or Florida, USA. This will be considered depending on staging factors of disease and how well your child is to travel. Your treatment centre will be able to discuss this with you including suitability for your child compared to treatment here in the UK using photon beam. We understand that this can cause added stress and will be very willing to discuss this with you further.

Chemotherapy

Chemotherapy is given together with surgery and or radiotherapy to treat the tumour. Treatment with anti-cancer drugs is used to destroy the cancer cells. It is usually given by injections and drips into a vein (intravenous infusion). The type of chemotherapy will vary according to whether the tumour has spread, if radiotherapy is to be used and the age of the child. Some children may need high dose chemotherapy with a stem cell transplant. If your child takes part in a clinical trial, the treatment is explained in more detail in a specific trial information sheet. You will be given an outline of the treatment in the form of a 'road map'.

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

Clinical trials may also involve research aimed at understanding more about the tumour's biology. We may ask you to allow us to do research on the tumour sample removed at surgery or on blood or CSF samples. Well run clinical trials have led to a significant improvement in the treatment of children with cancer. You can find more general information on clinical trials in the separate information sheet. If you are asked to consider entering your child into a clinical trial, we will give you specific information about the trial before deciding whether to take part. Your child's care will not be affected by your decision to take part in a clinical trial or not.

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 - these 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 and we usually

expect hair to regrow after treatment has stopped.

- 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 side effects of individual drugs, please see the specific drug information leaflets from Macmillan Cancer Support (details on page 7).

What is the outlook for my child?

Significant progress has been made in the management of children with medulloblastoma / PNET over the last 20 years. The majority of children with a localised tumour removed by the surgeon and who also have intensive chemotherapy and radiotherapy in a reduced dose to the brain and spine, are cured. Children who have PNET, an incomplete removal of tumour at initial surgery, or evidence of the tumour having spread, have a poorer outlook. The treatment of infants is difficult. This is because of the need to delay, avoid or reduce radiotherapy to the brain and spine, due to the damaging effects of radiation on the young developing brain.

What are the possible long term effects?

As medulloblastomas are more common 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 treatments. There is a small risk of developing a second cancer later in life.

All children are seen after treatment is finished in our long term brain tumour follow-up clinic. We will see your child at regular intervals indefinitely so that we can help with any long term effects of the treatment.

Other sources of information and support

General information on chemotherapy, hair loss, and side effects is available at The Royal Marsden. Please ask your CNS or contact the Help Centre:

The Royal Marsden Help Centre
Freephone: 0800 783 7176
Email: *patientcentre@rmh.nhs.uk*

Several organisations also offer support for children with brain tumours and their families:

Macmillan Cancer Support

Macmillan Supportline: **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

www.thebraintumourcharity.org/

The Brain Tumour Charity is a UK dedicated brain tumour 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.

CLIC Sargent for children with cancer

www.clicsargent.org.uk

CLIC Sargent is a UK charity for children and young people, and their families who provide clinical, practical, financial and emotional support to help them cope with cancer.

Brains Trust

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.

The Child Brain Injury Trust

www.childbraininjurytrust.org.uk

The Child Brain Injury Trust is a UK based charity which aims to support families of a child who has experienced an acquired brain injury, including a brain tumour. Practical advice and support is available, especially on how to manage the effects day to day and through education. Support can be provided face to face, phone and through email to parents and schools.

Contact details

Paediatric 24 hour advice line	0208 915 6248
If under Adult care	0208 915 6899
The Royal Marsden Macmillan Hotline	020 8915 6899
(available 24 hours a day, 7 days a week)	
Your Clinical Nurse Specialist Keyworker	0208 661 3805
Children's unit reception (for appointments)	0208 661 3602
Children's Day Unit	0208 661 3601
McElwain Ward (under 16 years)	0208 661 3611
Teenage Cancer Trust Unit (TCTU)	0208 915 6254

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
Freephone: 0800 783 7176
Email: patientcentre@rmh.nhs.uk

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

The Royal Marsden Cancer Charity

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