Medulloblastoma

What is a Medulloblastoma?

Medulloblastoma is a malignant tumour formed from primitive or poorly developed brain cells.

These tumours commonly arise in the posterior fossa of the brain, but are also found in other regions of the brain, including the pineal gland and cerebrum. Medulloblastomas are commonly referred to as a PNET (Primitive Neuro ectodermal tumour). They have the potential to spread (metastasise) to the spinal column via the cerebrospinal fluid (CSF), but rarely spread to other organs of the body.

Who gets a Medulloblastoma?

Medulloblastoma is the most common brain tumour of childhood, representing 20% of all childhood brain tumours. 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 occur in adults but are extremely rare.

What causes Medulloblastoma?

Like most brain tumours the cause of medulloblastoma is unknown. Research continues into finding a cause.
What are the signs and symptoms?
The symptoms are usually due to increased pressure in the skull (raised intracranial pressure).
Common symptoms include:
- Vomiting (most common), with or without nausea. Commonly occurs in the morning after waking up.
- Lethargy and irritability
- Headaches
- Clumsiness
- Difficulty with tasks like handwriting
- Gradual decline in school performance (impaired attention)
- Changes in personality and behaviour.
- Abnormal neck posture
- Abnormal gait (The way they walk)

If the tumour spreads to the spinal cord, the signs and symptoms may include:
- Back pain
- Difficulty walking
- Problems with bowel and bladder control.
The symptoms your child has will depend on the size and position of the tumour.

What tests will be performed?
It is necessary to find out as much as possible about the type, position and size of the tumour. These tests include:
- Neurological examination - to assess any effects the tumour has had on the nervous system.
- CT Scan and or MRI (Magnetic Resonance Imaging) of the whole brain and the spinal cord will be done to find the exact position and size of the tumour. Spinal imaging is necessary to rule out spread of disease of the spinal cord. (A full explanation of these test is included in the procedure section in this folder)
- Biopsy – A sample of the tumour will be taken when surgery occurs to confirm the exact type of tumour.
• Other tests will include a lumbar puncture to detect the presence of tumour cells in the cerebro-spinal fluid (CSF).

How do you treat a Medulloblastoma?

Results from the tests will enable your doctor to discuss the best forms of treatment.

Common treatments include:

• Surgery
• Radiotherapy
• Chemotherapy

Surgery

Medulloblastoma may block the ventricles (draining tubes) of the brain and cause a build up of CSF (hydrocephalus), which raises the pressure within the skull. If this has occurred before surgery can be performed this pressure may need to be reduced and a SHUNT may have to be inserted to drain off the excess fluid. (Information on shunts included further on.)

The aim of surgery is to remove as much of the tumour as possible without damaging the healthy brain tissue. As tiny (microscopic) tumour cells may remain after surgery it is followed up with radiotherapy and or chemotherapy.

Some tumours cannot be removed surgically as the risk of damage to surrounding brain cells is too high. If surgery is not possible your doctor will discuss other forms of treatment.

Following surgery for medulloblastoma, a CT or MRI scan will be done to determine if any tumour remains.

Radiotherapy

Radiotherapy is commonly used after surgery to destroy any remaining malignant cells. As medulloblastoma may spread through the CSF to the spinal cord, radiotherapy is given to the brain and spinal cord.

Because of the significant effects of radiotherapy on an infant’s developing brain, children under the age of four rarely undergo radiotherapy; they would receive chemotherapy following surgery.
Please ask for the radiotherapy section if you require further information.

Chemotherapy
Chemotherapy is given together with surgery and or radiotherapy to treat the medulloblastoma.

What is the prognosis?
Significant progress has been made in the management of children with medulloblastoma over the last 10 years. Intensive chemotherapy and reduced dose radiation to the brain and spine has seen a rise in the overall survival rate. Currently, there stands an 80% survival rate at 5 years from diagnosis in children that have had complete removal of the tumour at initial surgery, with no evidence of tumour spread to other parts of the central nervous system. Children, who have had an incomplete removal of tumour at initial surgery, or evidence of tumour spread, have a reduced survival rate. Currently this stands at 50 – 60 % survival at 5 years from diagnosis. The treatment of infants continues to be difficult. This is because of the delay in delivering radiation to the entire brain and spine.

Your treating doctor will discuss with you specific information relating to your child.

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, behavioural changes, possible learning problems and difficulties with coordination. Before treatment is commenced your doctor will discuss the possible long-term effects with you.
Shunts

A shunt is a system of tubing used to drain CSF (cerebrospinal fluid) from the ventricles or sub-arachnoid space into another area of the body. To understand why a shunt is used the basic anatomy and function of the skull, brain and CSF will be outlined.

The brain occupies most of the skull cavity and is surrounded by a cushioning layer of CSF. This fluid is primarily produced and circulated within the four interconnecting ventricles of the brain.

The ventricles contain a structure called the choroid plexus, which produces most of the CSF, approximately 500mls per day. This fluid continually circulates and contains many substances necessary for nourishment and normal function of the nervous system. CSF also provides a protective cushion within and around the brain.

CSF flows through the ventricles, and out three small openings in the fourth ventricle before it enters the sub-arachnoid space which surround the brain and spinal cord. The fluid then flows over the brain and spinal cord surface, and is eventually absorbed into the blood stream. The CSF is in a continual process of forming, circulating and then being absorbed. Under normal circumstances this is a delicate balance.

Anatomy of the skull, brain, ventricles and the circulation of the CSF (arrows). Note that CSF flows from lateral ventricle into third ventricle, aqueduct, and fourth ventricle and then escapes from the fourth ventricle into the subarachnoid space.

Diagram from Hydrocephalus- Patient Information Booklet- Medtronic.

Hydrocephalus will develop if the CSF cannot flow through the ventricular system, or if it is not absorbed into the blood. It will also occur if there is an over production of CSF related to a rare tumour known as a choroid plexus papilloma.
Types of Hydrocephalus

There are two types of hydrocephalus obstructive and non-obstructive.

Obstructive occurs when the CSF flow is obstructed within the ventricular system.

Non-obstructive occurs when CSF leaving the fourth ventricle is restricted in its flow over the surface of the brain, or if the sites of absorption are not functioning properly.
Treatment

The treatment for hydrocephalus is surgery to divert the CSF to the abdominal cavity, called a ventriculoperitoneal or VP shunt or to a chamber of the heart called the right atrium (a ventriculoatrial or VA shunt).

![Diagram of shunt system](image)

Illustrates the two most common shunts Ventriculoatrial (VA) and Ventriculoperitoneal (VP).

Diagram from Hydrocephalus- Patient Information Booklet- Medtronic

The shunt system usually contains two catheters and a one way valve. The catheter placed in the ventricle is called the proximal catheter and the catheter placed in the abdominal cavity is called the distal catheter. Both are attached to a one way valve used to regulate the amount, direction and pressure of CSF flow.
Some shunts include a reservoir, which can be used to flush the shunt or sample CSF. If this type of shunt is used, patients and families are discouraged from pressing the reservoir to “test” the shunt. This can be dangerous unless done under medical instruction.

**Shunt Surgery**

The surgery is a relatively short procedure and the length of time your child will stay in hospital will vary from patient to patient and your child’s surgeon will discuss this with you. It is necessary to have regular follow up appointments to check the shunt is working correctly. It may be necessary to have further CT Scans or MRI scan also to check that the shunt is functioning properly. Children can physically outgrow a shunt so it may be necessary from time to time to have revision surgery; your surgeon/doctor will discuss this with you.

**Possible complications**

It is important for you to be aware of the signs and symptoms that result as a complication of the shunt.

**Obstruction** – this is the most common complication experienced. Obstruction can occur from the catheter becoming plugged by brain or choroid plexus tissue or at the distal end by a loop of bowel, other structures or scarring.

The signs and symptoms of an obstruction are related to the increased pressure in the head. These will vary depending on the degree of obstruction.

- Partial or intermittent obstruction will result in periodic headache, nausea and vomiting, along with drowsiness, listlessness and decreased mental function.
- Complete obstruction will have rapidly developing headache, nausea, vomiting, blurring of vision, loss of coordination and deterioration of consciousness to the point of coma.
- In either case the doctor will perform tests to determine the degree of blockage and location and discuss the options for correcting the obstruction.

**Infection** – The risk of infection is present with any surgical procedure particularly when a foreign body (shunt) is implanted.

Observe for - any redness or swelling of wounds or along the path of the shunt system. Infections must be treated promptly.
Treatment will involve intensive antibiotic therapy and in many instances may require removal of the shunt.

**Overdrainage** – may produce a variety of signs and symptoms. Patients generally experience a headache that is worse when standing and reduced by lying down. Other symptoms may include nausea and vomiting, drowsiness and changes in vision.

**Precautions**

Care should be taken in relation to activities that could cause pressure on the shunt especially rough contact sports.

If at any time you are concerned please contact the hospital or your doctor.

**Bibliography**

Royal Children’s Hospital, Department of Haematology and Oncology Neuro-Oncology Parent Handbook.

Hydrocephalus – Patient Information Booklet 1996, Medtronic, Inc.

**Further Information**

Further information can be found in the following web sites and patient leaflets or in the section ‘Sources of more information on cancer’ section in the folder.

www.cancerbacup.org.uk/info/medulloblastoma.htm

www.rch.unimelb.edu.au/haem_oncology/pages/braintumours.html

A Patient’s Guide to Understanding Brain Tumours 2000, from Schering Oncology Biotech

www.stjude.org/diseasestudies/medulloblastoma_PNET.html