

Ependymoma (children)

Ependymoma is a type of brain tumour found in both children and adults although it is much more common among children than it is adults. Ependymoma is the third most common type of child brain tumour (following astrocytoma and medulloblastoma).

The information in this fact sheet gives an overview of ependymoma in children and answers some of the questions you may have about this type of tumour.

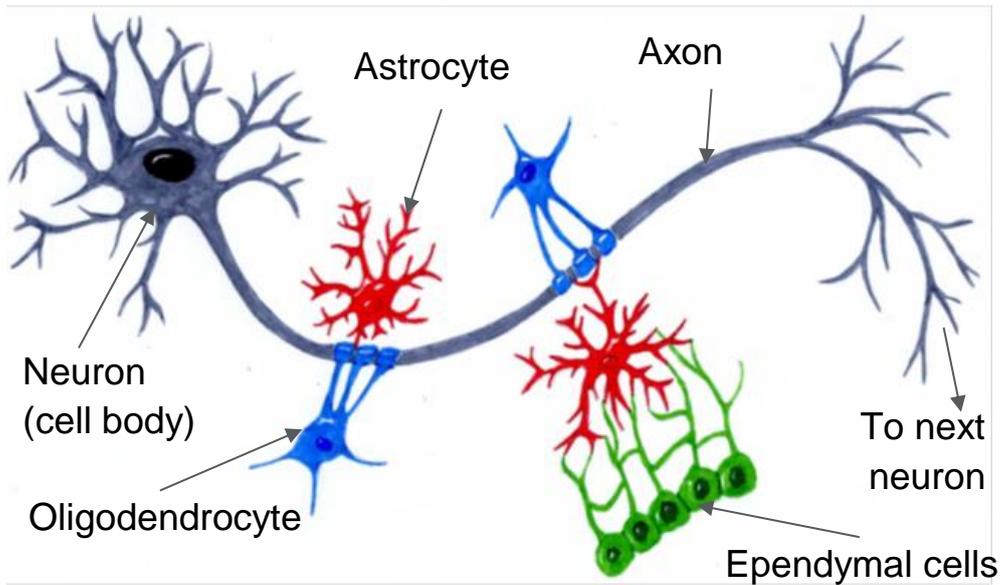
In this fact sheet:

- What is an ependymoma?
- Types of ependymoma
- What are the symptoms of ependymoma?
- How is an ependymoma diagnosed?
- How are ependymomas treated?
- How often do ependymomas occur?
- Further information

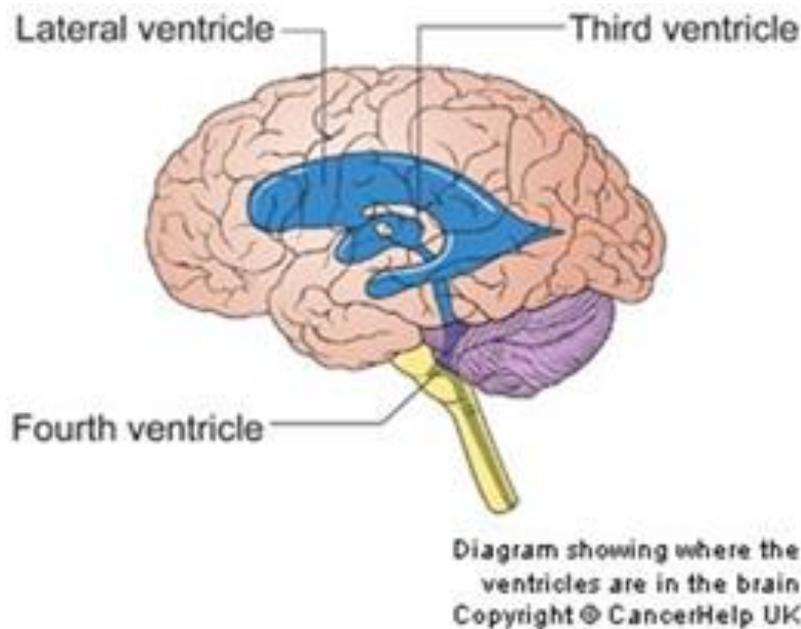
What is an ependymoma?

Ependymoma is a type of glioma (a tumour arising from glial cells). While neurons are responsible for the transmission of 'messages' through our central nervous system, glial cells support and protect neurons by providing them with oxygen and nutrients, and by removing dead cells. Glial cells are much smaller than neurons and we have many more glial cells than neurons.

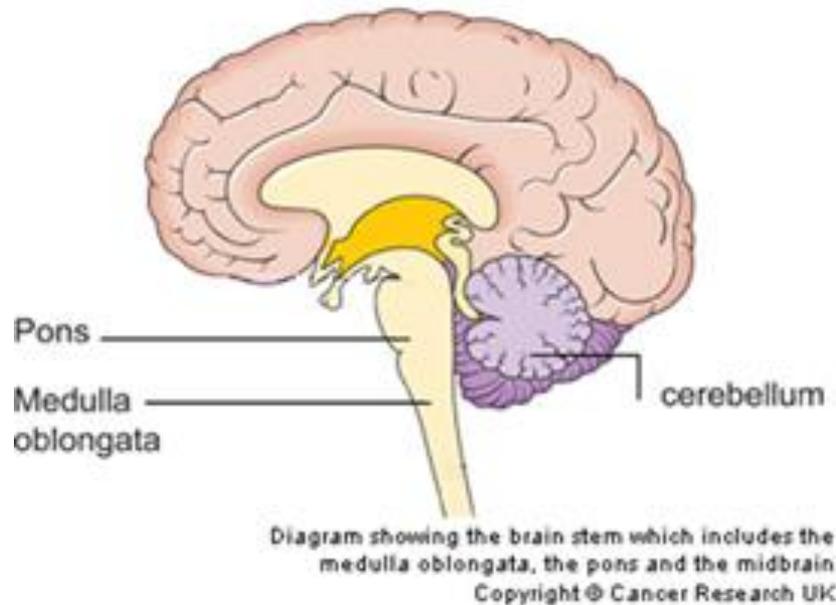
There are different types of glial cell, each of which plays a different role in supporting the neurons. The main types are astrocytes, oligodendrocytes and ependymal cells.



Ependymomas arise from ependymal cells. Ependymal cells are found lining the ventricles of the brain (the fluid-filled spaces in and around the brain). Two-thirds of the cerebrospinal fluid (CSF) surrounding, protecting, and nourishing the brain is produced by ependymal cells.



The majority of childhood ependymomas (around 60-70%) are located in the 'posterior fossa' or hindbrain, which is the area in the lower part of the brain containing the cerebellum and parts of the brainstem (pons and medulla).



Types of ependymoma

Ependymoma tumour cells don't always appear identical under the microscope and often display differences in behaviour. This section identifies the four subtypes of ependymoma and the differences between them.

Like most other types of brain tumour, the World Health Organization (WHO) classifies the subtypes of ependymal tumour (listed below) using a numerical grading scale. In general terms, a lower grade of 1 usually means that the tumour cell grows relatively slowly, is less likely to spread and looks substantially similar to a healthy cell under the microscope. On the other hand, the higher the grading of a tumour cell (2-3-4), the less it shares with healthy cells in terms of appearance, the faster it grows, and the more likely it is to spread to other parts of the brain or spinal cord. Listed below are the different subtypes of ependymoma along with their WHO grade:

- **Myxopapillary Ependymomas (Grade 1)** – This type of ependymoma is relatively uncommon in children and occurs more often in the lower part of the spinal column rather than the brain. There are two main types of glioblastomas: primary (sometimes called 'de novo' meaning 'from new') and secondary.
- **Subependymomas (Grade 1)** – Subependymomas are also relatively uncommon in children and appear more often near a ventricle.
- **Ependymomas (Grade 2)** – Ependymomas are the most common type of ependymal tumour and usually appear close or within the ventricular system in the posterior fossa.

- **Anaplastic Ependymoma (Grade 3)** – Anaplastic ependymomas are the fastest growing type of ependymal tumour and also commonly originate in the posterior fossa.

While the classification presented above helps in understanding some basic variations between ependymal tumours, the appearance of ependymoma cells under the microscope doesn't always fit or predict their behaviour and possibility of recurrence. For this reason, it is broadly accepted by the medical community that more large-scale studies are needed in order to identify the exact factors determining ependymal tumour behaviour and to subsequently revise the existing grading scheme.

What are the symptoms of ependymoma?

The signs and symptoms, that a child with an ependymoma may display, depend on a number of factors, including the age of the child, the location of the tumour and its size.

An ependymoma may cause two main types of symptoms: general and focal.

General symptoms

General symptoms refer to those that might be caused by a mass in the brain, regardless of where it is located. General symptoms of tumour growth in children and adults are associated with a build-up of pressure in the head. This build-up of pressure is either caused because the tumour itself is taking up space in the skull or because of hydrocephalus (when the tumour is blocking the drainage of cerebrospinal fluid). The following are common general symptoms of a brain tumour:

- Enlargement of the head (in infants)
- Persistent headache (especially in the morning)
- Recurrent vomiting
- Seizures
- Changes in vision
- Sleepiness
- Cognitive difficulties (unexpected difficulties thinking, concentrating, remembering or thinking).

(For more information on brain tumour symptoms in children visit www.headsmart.org.uk/home/)

Focal symptoms

Focal symptoms refer to symptoms caused specifically by the position of the tumour within the brain. As mentioned already, the majority of child ependymomas occur along ventricles (liquid filled spaces) and within the posterior fossa or hindbrain area of the brain. The part of the ventricular system within the hindbrain area is known as the 'fourth ventricle'.

Depending on where in the fourth ventricle the tumour is located, a child might display one or more of the following symptoms:

- 'Wry neck' with the head appearing tilted and twisted ('torticollis')
- Loss of balance ('ataxia')
- Problems articulating speech ('dysarthria')
- Difficulty swallowing ('dysphagia')
- Making overly clumsy movements ('dysmetria')

If your child displays one or more of the general or focal symptoms, consult a doctor who may run further diagnostic tests to determine their cause.

How is an ependymoma diagnosed?

If your doctor (GP or A&E doctor) suspects your child has a brain tumour, s/he will refer him/her to a specialist - a neurologist (specialists in brain and nerve disorders), or an oncologist (specialist in treating cancer). The specialist will ask questions about your child's health and give them a physical examination.

They will also test their nervous system (called a neurological examination). This involves looking at your child's vision, hearing, alertness, muscle strength, co-ordination, and reflexes. They will also look at the back of their eyes to see if there is any swelling of the optic disc (the optic disc is where the optic nerve from the brain enters the eye). Any swelling is a sign of raised pressure inside the skull, which could be a sign of a brain tumour.

Your child will then have one or more further tests. This may include a blood test to check for certain 'markers' in the blood that some tumours can cause. They will also need an MRI (magnetic resonance imaging) or CT (computerised tomography) scan to confirm whether a brain tumour is present. (For information about these scans, see the Scans fact sheet).

If from a scan, a tumour is identified, a resection (biopsy) will be planned to remove as much of your child's tumour as possible for diagnosis (for testing).

A resection to remove a brain tumour is a complex operation which takes several hours. Any risks will be explained to you by your child's surgical team. If possible, the neurosurgeon will attempt to remove the whole tumour during the resection.

In both cases, cells from the tumour will be analysed in a laboratory by a neuropathologist (See The Multi-Disciplinary Team (MDT) fact sheet). The neuropathologist will examine the cells under a microscope and make a diagnosis based on the characteristic tissue patterns of ependymoma. Also, the tumour will be divided into one of the four different sub-groups (types). It is important that a detailed diagnosis of the exact tumour type is made as this will allow your medical team to determine the best course of treatment for your child.

How are ependymomas treated?

The development of effective treatments tailored to manage specific types of tumour usually depends on how much is known about that type of tumour; about the factors which contribute to its appearance and behaviour.

Knowledge about these factors helps with the development of drugs which can target the tumour effectively. There is ongoing research being carried out in order to understand more about the causes of ependymoma in order to develop more targeted treatments than the ones currently offered.

Surgery

Surgery is the first line of treatment for ependymomas. Through surgery, the neurosurgeon will aim to remove as much of the tumour as possible. After surgery, the doctor will create a treatment plan of consisting of radiotherapy or radiotherapy with chemotherapy, based on the amount of tumour removed, the child's age and whether the disease has spread. (For more information on Biopsy and Surgery see the Neurosurgery for children fact sheet or watch the Jake informational animation at: www.bit.ly/Jake-Neurosurgery)

Radiotherapy

Radiotherapy, or radiation treatment, uses controlled doses of high energy beams to destroy tumour cells whilst causing as little damage as possible to surrounding cells. Radiotherapy may be used where surgery isn't possible, or after surgery to kill any remaining ependymoma cells. It can also be used to prevent a tumour from returning or to slow down its growth.

Studies have shown that radiotherapy can be effective in managing ependymomas. However, there are short and long-term risks associated with

treating children with radiation, especially those younger than three years old. The risks and benefits for each child with ependymoma have to be weighed before deciding if radiation therapy should be given in any form. (For more information see the Radiotherapy for children fact sheet or watch the Jake informational animation at: www.bit.ly/Jake-radiotherapy)

Chemotherapy

Chemotherapy is a treatment which involves taking drugs that have been developed to kill tumour cells. Chemotherapy drugs are taken either orally in the form of a pill or intravenously using an injection or drip, and a treatment cycle is usually spread over a set period of time.

Results of clinical trials on the effectiveness of individual chemotherapy drugs for childhood ependymoma have been mixed with some drugs appearing ineffective in targeting ependymomas and others reporting effectiveness in a limited percentage of cases. There is ongoing research on the possible effects of different combinations of chemotherapy drugs and radiotherapy in treating childhood ependymomas. (For more information see the Chemotherapy for children fact sheet or watch the Jake informational animation at: www.bit.ly/Jake-chemotherapy)

How often do ependymomas occur?

Ependymomas are the third most common brain tumour in children and individuals under the age of 18, accounting for around 10% of paediatric brain tumours and close to 2% of all paediatric cancers. Half of all childhood ependymomas arise in children younger than 5 years of age and around one third in children under 3 years.

Further Information

To access all our Jake informational animations for children visit:
www.bit.ly/Introducing-Jake

For more information about childhood brain tumours and symptoms visit:
www.headsmart.org.uk/home/

What if I have further questions?

If you require further information, any clarification of information, or wish to discuss any concerns, please contact our Support and Information Team.

Call 0808 800 0004 (free from landlines and most mobiles including 3, O2, Orange, T-mobile, EE, Virgin and Vodafone)

- Email: support@thebraintumourcharity.org
- Join our closed Facebook group:
bit.ly/supportonfacebook

About us

The Brain Tumour Charity makes every effort to ensure that we provide accurate, up-to-date and unbiased facts about brain tumours. We hope that these will add to the medical advice you have already been given. Please do continue to talk to your doctor if you are worried about any medical issues.

The Brain Tumour Charity is at the forefront of the fight to defeat brain tumours and is the only national charity making a difference every day to the lives of people with a brain tumour and their families. We fund pioneering research to increase survival, raise awareness of the symptoms and effects of brain tumours and provide support for everyone affected to improve quality of life.

We rely 100% on charitable donations to fund our vital work. If you would like to make a donation, or want to find out about other ways to support us including fundraising, leaving a gift in your will or giving in memory, please visit us at www.thebraintumourcharity.org, call 01252 749043 or email fundraising@thebraintumourcharity.org

About this fact sheet

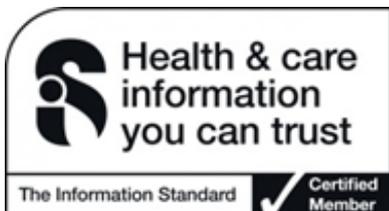
This fact sheet has been written and edited by The Brain Tumour Charity's Support and Information Team. The accuracy of medical information has been verified by a leading health professionals specialising in neuro-oncology. Our fact sheets have been produced with the assistance of patient and carer representatives and up-to-date, reliable sources of evidence. If you would like a list of references for any of the fact sheets, or would like more information about how we produce them, please contact us.

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Your notes



Hartshead House
61-65 Victoria Road
Farnborough
Hampshire
GU14 7PA
01252 749990
enquiries@thebraintumourcharity.org
www.thebraintumourcharity.org

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