

Childhood brain tumours

Our bodies are made up of billions of cells. Normally, these cells reproduce and repair themselves in a controlled way and do not cause us any problems. If for some reason this process gets disrupted, the cells can begin to grow in an uncontrolled way, creating a lump of cells called a tumour.

A tumour that grows in the brain is called a brain tumour. Tumours can also grow along the spinal cord. Brain and spinal tumours are Central Nervous System (CNS) tumours. There are over 100 different types of brain and spinal tumour. This fact sheet explains some of the terms you may hear if your child receives a diagnosis of a brain tumour.

In this fact sheet:

- What is a brain tumour?
- How common are childhood brain tumours?
- How is a brain tumour diagnosed?
- What causes brain tumours?
- How are childhood brain tumours treated?

- What are the side effects of a brain tumour?

The diagrams in this fact sheet were taken from the patient information website of Cancer Research UK: www.cancerresearchuk.org

What is a brain tumour?

A brain or CNS tumour is an abnormal growth caused by cells dividing in an uncontrolled way. There are over 100 different types of brain and spinal tumour and they are usually named according to the types of cell they start to grow from and/or parts of the brain they grow in. A primary brain tumour begins somewhere in the brain - it has not spread from elsewhere in the body. A secondary brain tumour (known as a metastasis) has begun somewhere else in the body (for example, the kidney or skin) and has spread to the brain.

(For more information about tumour types, please see the sections at the end of this fact sheet)

How common are childhood brain tumours?

Childhood brain tumours are relatively rare. Around 500 children and young people in the UK are diagnosed with a brain tumour each year. This means that most times the symptoms your child is showing **will not be due to a brain tumour**. However, it is important to be aware of the symptoms, so you can go to your doctor if you are concerned.

How will I know if my child has a brain tumour?

Symptoms of brain tumours vary from child to child. Symptoms can also depend on exactly where in the brain the tumour is and can often mimic those of other, relatively minor childhood illnesses. The presence of a symptom does not necessarily mean that your child has a brain tumour.

Common symptoms of childhood brain tumours include:

- Persistent vomiting/feelings of nausea (over a two week period)
- Recurring headache (over a four week period, particularly on waking)
- Abnormal eye movements
- Fits or seizures
- Behaviour change
- Abnormal balance/walking/co-ordination
- Blurred/double vision
- Abnormal head position (such as a head tilt)

- Delayed or arrested puberty (puberty that doesn't start or starts, but doesn't progress as expected)

If your child has one or more of the above, you should take them to see a GP, explaining your worries about a brain tumour being present. If they have two or more, ask for an urgent referral. An urgent referral means that your child will be given an appointment with a specialist who can further investigate the cause of their symptoms.

You will find much more information about childhood brain tumours on our 'HeadSmart – be brain tumour aware' website:

headsmart.org.uk.

HeadSmart is our UK-wide campaign that aims to reduce the time it takes to diagnose children and young people with a brain tumour and so achieve better outcomes - saving lives and reducing long-term disability.

We have pocket-sized symptoms cards that list common signs and symptoms of childhood brain tumours that you can take with you to your family GP if you are concerned about your child. These are available to view on the HeadSmart website.

You can also get a link to the cards sent directly to your smartphone by texting SMART to 81400 or we can post a card to you. Call our Support and Info Line on **0808 800 0004** or email **support@thebraintumourcharity.org** to request a card or for more information.

How is a brain tumour 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 or neurosurgeon (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. They may have a blood test to check for certain 'markers' in the blood that some tumours can cause. These could be changes in the levels of certain hormones. 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 for children fact sheet).

If, following the scan, a tumour is found, a biopsy (small sample of the tumour) may be taken from your child's tumour. It is important to realise that a biopsy is an operation that takes several hours. Any risks will be explained to you by your child's surgical team.

Alternatively, if possible, the resection (surgical removal) of the whole tumour will be undertaken.

In both cases, cells from the tumour will be analysed in a laboratory, so that health professionals can give a more detailed diagnosis of the exact tumour type. This will allow them to determine the best course of treatment for your child.

'Benign' or 'malignant'

Your child's brain tumour may be referred to as 'benign' or 'malignant'. A benign brain tumour is relatively contained and is unlikely to spread to other parts of the brain. It tends to be slow growing and there is less chance of it returning if it can be completely removed.

A benign brain tumour can still be serious, however, due to the limited space capacity of the skull or difficulties removing tumours from certain parts of the brain.

A malignant brain tumour is faster growing. It is far more likely to spread to other parts of the brain or spinal cord and may come back, even if treated. Brain tumours very rarely spread to other parts of the body. Malignant brain tumours cannot usually be treated by surgery alone - other treatments such as radiotherapy and/or chemotherapy may also be used.

Grades

The terms benign and malignant may not be used in relation to your child's brain tumour. Doctors could instead refer to gradings. The grading system goes from 1 to 4. A Grade 1 tumour is the slowest growing while a Grade 4 is the quickest growing, most aggressive kind. Within a tumour, it is possible to have cells of different grades (for example, some grade 1 cells and some grade 2 cells) but the grade the tumour is given as a whole will be determined by the highest grade of cells found.

It is important to know that brain tumours in children rarely change over time or increase in grade. This is more often seen

in adults.

Gradings can be quite difficult to understand. Your child's doctor will explain what it means for your child. Do ask questions if you are not clear.

What causes brain tumours?

Very often, the answer is that we just do not know. This can be one of the most difficult things to accept as a parent as it can leave you feeling helpless. There has been much in the news about the possibility of mobile phones and power lines causing brain tumours, but research remains inconclusive and, at present, there is no clear link between exposure to mobile phones or power lines and brain tumours.

Brain tumours are nobody's fault. There is no research that shows that anything you may have done during pregnancy or in your child's early infancy could have caused your child's brain tumour. In addition, a tumour that has gone undetected for some time is not your fault - brain tumours can be difficult to spot as their symptoms can be very similar at first to many common childhood ailments.

The risk factors that we know about

Genetics

According to Cancer Research UK, genetics (inheriting a gene or genes that make you more likely to get a brain tumour) are thought to account for around 5% of brain tumours. The following factors may affect your child's risk of developing a brain tumour:

- Certain genetic conditions (including neurofibromatosis type 1 and 2, tuberous sclerosis, Li-Fraumeni syndrome, Von Hippel-Lindau syndrome or Turner syndrome) may increase your child's risk of developing a brain tumour.
- If an immediate family member (parent or sibling) has a tumour of the CNS your child may have a slightly higher chance of developing a brain tumour, although the risk is still very low.

Radiation

We know that radiation does increase the risk of a brain tumour developing. The risk of developing a meningioma or glioma (types of brain tumour that you can read more about further on in this fact sheet) is higher in children who have had radiotherapy to the head, particularly before they reach the age

of five. Due to the known risk of learning difficulties, health professionals try to avoid irradiating a young child's brain. Radiotherapy is not given without very careful consideration and your child would not receive radiotherapy unless the expected benefits outweighed any potential risks to them.

How are childhood brain tumours treated?

Several factors influence the decision as to which treatment will best help your child. A team of specialised health professionals will consider your child's individual diagnosis and take account of factors, such as the size and location of the tumour, the type of tumour your child has and how quickly it is growing. They will also consider your child's age and their general health.

Surgery will often be used to remove as much of the tumour as possible. This will help to reduce pressure on the brain caused by the impaired flow and accumulation of the CSF, the tumour itself or by the brain's reaction to its growth (cerebral oedema). It is increased pressure that can cause some of the symptoms.

However, depending on where in the brain the tumour is, surgery is not always possible or necessary. Sometimes, it would be too risky to operate as the tumour may be very close to, or wrapped around, an important part of the brain, such as the brain stem, and the benefits of surgery would be outweighed by the dangers. In other cases, such as with very slow growing, low grade brain tumours, problems with increased pressure may not develop, so your child may not need surgery straight away or not even at all. (*For more information, see our Neurosurgery in children fact sheet*)

Other treatments, such as **radiotherapy** (generally given only to children who are at least three years old) and/or **chemotherapy** may be used on their own, in combination or after surgery to try to remove any remaining tumour cells. (*For more information, including information about duration of treatment, please see our Radiotherapy for children and Chemotherapy for children fact sheets*).

A further type of treatment, that your child may have, is **Proton Beam Therapy** (PBT). PBT is a type of radiotherapy that uses a beam of positively charged subatomic particles (protons) to target the tumour in a very precise way, reducing the harm to healthy tissue. PBT is not suitable for all types of brain tumours. It works best for smaller tumours and those where the margins are clearly defined. It is also not currently available in the UK for use in brain tumours. PBT can, however, be received outside the UK, through the NHS, if it would be suitable for your child.

The Department of Health has announced two Trusts in England that have been chosen to develop PBT centres - The Christie NHS Foundation Trust Hospital in Manchester and University College London Hospitals NHS Foundation Trust. Given the complex nature of the treatment and facilities, PBT won't be fully available in England until 2017. Until then, the NHS will continue to fund patients to receive treatment abroad.

It is important to note that there are specific referral criteria for PBT and if your child does not meet these criteria they will not be referred on the NHS. *(For further information, see the Proton Beam Therapy fact sheet).*

It is important to know that your child's medical team will tailor your child's treatment to provide the treatment that is the best for your child. This could mean that you meet other families whose children have the same tumour but who are receiving different treatments and the reasons for this could be due to several factors. If you are worried or concerned at all, you should speak to your child's medical team who will be able to help you understand the treatment decisions that have been made. If you feel your child is not getting the correct treatment you can ask for a second opinion. More information about this can be found on the NHS Choices website:

www.nhs.uk/chq/Pages/910.aspx

Will my child have learning difficulties as a result of their brain tumour?

Children who have had a brain tumour may experience learning difficulties. This could be because of the tumour itself, the impact of treatment, or the fact that they have had to take time out of school and therefore miss lessons. Unfortunately, it is generally the case that the younger a child is when they have a brain tumour, the higher the likelihood of them needing specialist educational input to support their learning. *(For further information, see the Learning difficulties and childhood brain tumours fact sheet)*

Generally, radiotherapy is given only to children who are at least three years old. A very young brain (under three years) is less developed and changing fast and so more likely to experience long-lasting damage from radiation. This poses a significantly increased risk of long-term social and educational learning difficulties. However, radiotherapy may be given to under threes in certain circumstances where the benefits are felt to outweigh the risks.

If your child receives radiation, you may not be aware of difficulties straight away and they may only become apparent as your child gets older. *(For further information, see the Radiotherapy for children fact sheet)*

What other long-term effects might my child have?

Long-term effects from a brain tumour vary from child to child and depend on the exact location of the tumour within their brain and the therapy given. Your child's consultant will be able to talk you through the possible long-term effects they might experience. You should be able to ask any questions of them that you would like to.

Some possible long-term impacts include:

- **Sight problems** - Brain tumours are the biggest preventable cause of blindness in children. Sight loss may be partial, for example, children may experience double or blurred vision, or their sight loss may be full. Sight loss depends on the area of the brain where the tumour grows and also the areas that receive radiation. This side-effect may develop over years. As a general rule, radiologists plan radiation to avoid the optic nerves if possible.
- **Impacts on puberty and fertility** - Treatments, such as radiotherapy and chemotherapy, can delay puberty and affect fertility. This can be addressed with an artificial hormone replacement. Girls who have radiotherapy to the head as a child may begin puberty earlier. However, medication can be used to stop this until your daughter is the right age to go through puberty. Speak to your child's medical team.
- **Impacts on growth** - If your child receives radiotherapy to, or near, the pituitary gland (which makes and releases chemicals responsible for many of the body's functions, including growth), growth may be slowed. This may be addressed with injections of an artificial growth hormone. Your child's growth may also be affected if your child receives radiotherapy to their spine as this may prevent the spine growing normally as your child develops.
- **Physical problems** - Some children who have had a brain tumour will be left with a long-term physical effect, such as weakness in a limb (hemiplegia), or difficulties with balance. Physiotherapy is one possible treatment that may help children to adapt.
- **Emotional difficulties** - Children who have had a brain tumour may experience emotional difficulties. This could be due to a number of factors, including changes to areas of the brain that control emotions, the emotional impact of a serious illness and also of the treatment itself. In addition, your child may experience emotional impacts from social

changes, loss of contact with their friends and being different from their friends, as well as fears of their tumour returning and of dying.

Coping as a family

Coping with the diagnosis of a childhood brain tumour can have a huge impact on you, as parents/carers, and on the rest of your family. Although centres often have several children with brain tumours on treatment at any one time, it could be that although your child is treated on a children's ward, they may be the only child on the ward with a brain tumour. This could feel lonely for them and for you.

The Brain Tumour Charity has a variety of services to give help if you or your child need it, including a dedicated Children and Families Worker, who can offer both practical and emotional support to parents, children and siblings. If you have questions or would like to talk to a member of our Support and Information team, please phone **0808 800 0004** or email **support@thebraintumourcharity.org**

We run Family Days, which provide children affected by brain tumours the opportunity to meet other children in similar situations in a social situation.

For further information, visit our website:

thebraintumourcharity.org/familydays

You may also find it helpful to read or contribute to our online discussion forums (listening rooms), which offer a safe space to gain peer support. We also have one specifically for young people under 18.

thebraintumourcharity.org/forums

Brain tumour types in children

Brain tumours in children are rare, but they do happen. There are many different types of brain tumour and their names can sound long and complicated. As a general rule though, tumours are named according to the type of cell they start from and/or where in the brain they are located. The following is not an exhaustive list.

By cell type

Gliomas

Throughout the brain and spinal cord we all have nerve cells called 'neurons', which transmit 'messages' (electrical and chemical signals). Surrounding our neurons are cells called glial cells. Glial cells provide our neurons with

oxygen and nutrients and remove dead cells, supporting and protecting the neurons. Glial cells are much smaller than neurons and we have many more glial cells than neurons. Gliomas are brain tumours that begin in these glial cells.

As there are different types of glial cells, including astrocytes, oligodendrocytes and ependymal cells, gliomas can also be divided into different types.

Types of glioma include:

- **Astrocytomas** (arising from astrocytes)
- **Ependymomas** (arising from ependymal cells)
- **Oligodendroglioma** (arising from oligodendrocytes)

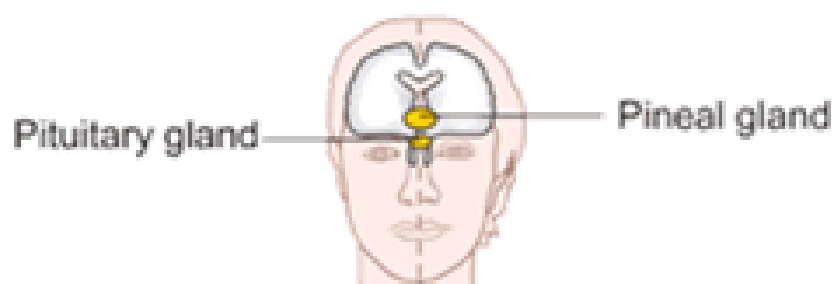
You may therefore, for example, hear a tumour referred to as an 'astrocytoma' or a 'glioma' (with astrocytoma being a more specific description of the tumour).

Primitive neuroectodermal tumours (PNETs)

PNET tumours develop from cells that are left over from the earliest stages of our development, while we are still growing in our mother's womb as an embryo. The 'neuroectoderm' is the part of the embryo that goes on to develop into the brain and spinal cord, but some cells do not develop and specialise the way other cells do and so they appear "primitive".

PNET tumours include:

- **Medulloblastomas** (which develop in the cerebellum)
- **Pineoblastomas** (which develop in the pineal region of the brain)
- **Central nervous system (CNS) PNET** (which develop in the upper parts of the brain). These used to be called non-pineal supratentorial PNETs.



The tentorium

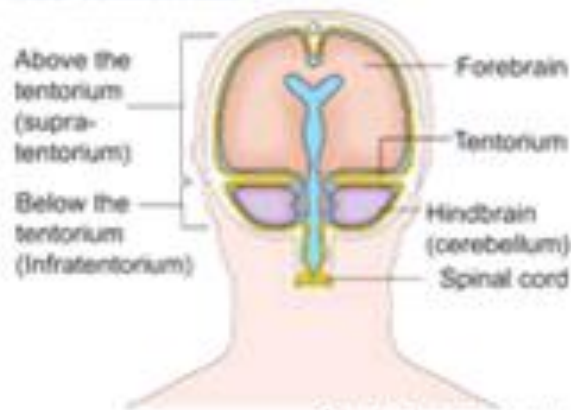


Diagram showing where the tentorium is in the brain
Copyright © Cancerhelp UK

As you can see, PNET tumours often also use the other means of naming tumours - their location in the brain.

By location in the brain

Brain tumours are also often given names which refer to where they are in the brain.

Posterior fossa tumours

'Fossa' means cavity or pit, so the posterior fossa is the cavity inside the posterior or back of the skull.

It contains, or is near, structures including the cerebellum (involved in balance, movement and co-ordination), the medulla (involved in involuntary functions such as heart rate and breathing) and the brainstem (involved in functions such as breathing, blood pressure and digestion).

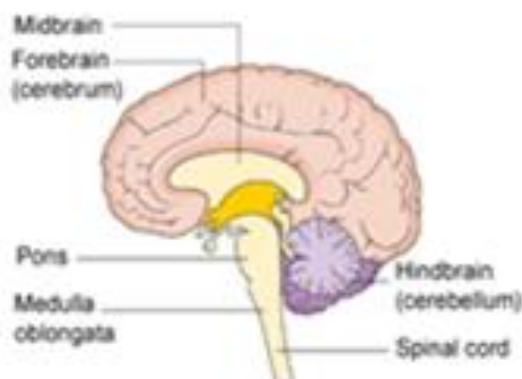


Diagram showing the brain stem which includes the medulla oblongata, the pons and the midbrain
Copyright © Cancerhelp UK

The hindbrain (cerebellum)

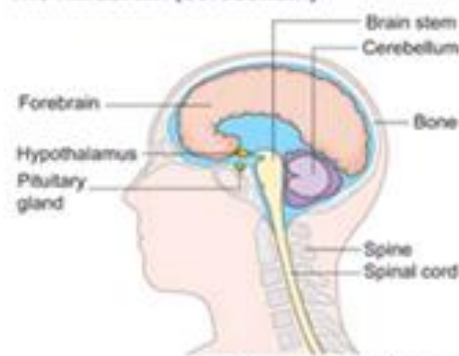


Diagram showing the parts of the brain
Copyright © Cancerhelp UK

Examples of tumours in the posterior fossa include:

- **Cerebellar pilocytic astrocytoma** - originates in astrocyte-type glial cells situated in the cerebellum. Pilocytic means the cells are elongated and look hair-like.
- **Medulloblastoma** - develops within the cerebellum
- **Ependymoma** - develops in the ependymal cells, which are the glial cells that form the lining of the ventricles (fluid-filled cavities) and tiny passageways of the brain and spinal cord. In children these tumours occur most often in the ependymal cells of the cerebellum, i.e. within the posterior fossa.
- **Brainstem glioma** - such as Diffuse Intrinsic Pontine Glioma (DIPG). These gliomas can develop in various parts of the brain because glial cells are found in all areas of our brain, so can occur in the brain stem.

Sometimes the term for the location within the brain will be used in addition to the name derived by the cell type e.g. posterior fossa ependymoma.

Further information about these tumour types is given in the 'Common childhood brain tumours' section below.

Cerebral hemisphere (cerebrum/forebrain) tumours

Another common site for brain tumours is in one of the cerebral hemispheres (also known as the cerebrum or forebrain) or in the spinal cord.

As glial cells are found in all areas of our brain, gliomas and their different types can also be found in the cerebral hemispheres. Similarly, primitive neuroectodermal (PNET) cells can also occur in the cerebral hemispheres.

Tumours within the cerebral hemispheres include:

- **Astrocytoma**
- **Ependymoma**
- **Oligodendroglioma**
- **Central nervous system PNET**

Central brain tumours

Other tumours are located in the central part of the brain and include:

- **Pineoblastoma** - these primitive tumour cells develop in the pineal region, commonly found near the base of the cerebral hemispheres in the pineal region and the area above the pituitary gland).

- **Craniopharyngioma** - these grow near the base of the cerebral hemispheres and arise on the stalk of the pituitary gland.
- **Choroid plexus carcinoma** - the choroid plexus is a structure within the cerebral ventricles that produces the cerebro-spinal fluid (CSF), the fluid within the brain.
- **Germ cell tumours** - germ cells are primitive cells that develop into the reproductive system; occasionally they can occur in the brain, where they are most commonly found near the base of the cerebral hemispheres in the pineal region and the area above the pituitary gland.

For further information about areas of the brain, see The human brain fact sheet.

Common childhood brain tumours

Glioma

Around half of all childhood brain tumours are a type of glioma (derived from the various glial cells). Of these, the largest subgroup is astrocytoma and these constitute over two-fifths (43%) of all brain and spinal tumours in children.

Astrocytomas

These tend to be slow growing (73% are low grade) and are diagnosed throughout childhood with no strong pattern by either sex or age.

Cerebellar pilocytic astrocytoma is the most common subtype of low grade glioma. This type of brain tumour occurs in the cerebellum (the structure in the posterior fossa that is responsible for balance, movement and co-ordination) and originates in the star-shaped astrocytic cells. Cerebellar pilocytic astrocytomas tend to be cystic, which means that they are associated with a single or several cysts. The vast majority (around 80%) of astrocytomas occurring in the cerebellum are low grade (slow growing). They are typically operable and the prognosis tends to be very good.

However, approximately 10-15% of childhood brain tumours in the UK are a type of astrocytoma or brain cell glioma called a Diffuse Intrinsic Pontine Glioma (DIPG). DIPG is a fast growing, malignant type of brain tumour that originates from astrocytic cells in the brain stem. They often appear in children aged around six years old. These tumours grow very fast within a very sensitive part of the brain so can be quite large and cause significant symptoms to show within a few days of their onset - unsteadiness, squint and swallowing symptoms are common.

Ependymomas and Choroid plexus tumours

Ten per cent of childhood brain and spinal tumours are defined as ependymoma and choroid plexus tumours. Ependymomas commonly occur within the ventricles of the brain or the posterior fossa and can block the spinal fluid from circulating and draining. Similarly choroid plexus tumours grow within the ventricles. The first symptoms of an ependymoma or choroid plexus tumour may therefore be pressure headaches, particularly in the mornings. They occur most often in one year olds, where the signs of the raised intracranial pressure may be vomiting and lethargy (drowsiness), rapidly increasing head size and an inability to look upwards.

PNET tumours

The second most frequent subgroup consists mostly of the primitive neuroectodermal tumours (PNETs). About 20-25% of childhood brain tumours are PNETs, which are malignant tumours. PNETs occur most frequently in younger children and incidence decreases with age with more than half of all PNETs being diagnosed in children less than 10 years old. Nearly three-quarters (73%) of PNETs are medulloblastomas.

Medulloblastomas

These are the most common type of malignant tumour in children. They make up around 20% of all childhood brain tumours. They are more common in boys than girls and are most commonly diagnosed at around the age of 5 years.

They develop in the cerebellum, a structure involved in co-ordination and movement, but may spread to other parts of the brain and spinal cord. Treatment depends on many factors (including the age of your child and the size and location of their tumour) but may involve surgery, radiotherapy and chemotherapy.

Other tumours

Craniopharyngioma

These are more common in children and young people than adults and account for 8% of childhood brain tumours. They do not usually spread, but are near important structures in the brain and can cause problems as they grow. They can cause changes in hormone levels and problems with eyesight. Children with craniopharyngioma can experience weight gain and growth problems.

From these descriptions you can see that a large proportion (around 60%) of childhood brain tumours start in the posterior fossa.

Rarer childhood brain tumours

Astrocytoma (high grade)

In children, 20% of astrocytomas are high grade and are classified as either anaplastic astrocytomas (grade III) or glioblastoma (sometimes referred to as GBM). These are malignant tumours. Glioblastomas are the quickest growing type of brain tumour (also sometimes called a grade 4 astrocytoma). Glioblastomas are rare in children.

Oligodendrogliomas

These tend to occur in the cerebrum (which covers much of the brain) in the frontal or the temporal lobes. They are far less common in children than astrocytomas and ependymomas.

Meningiomas

These tend to start in the membrane covering the brain, called the meninges. They are slow growing tumours. They are rare in children, but may occur in children who have neurofibromatosis type 2 - a genetic condition that causes tumours to grow along your nerves. They may be associated with radiotherapy given at a young age. The usual therapy is surgery.

This list of tumours is not exhaustive and there are other rare brain and spinal tumours in childhood. The list of names for the tumours is expanding due to continuing developments in the understanding of the microscopic structure of tumour cells.

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, which includes a dedicated Children and Families Worker:

- 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.

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 thebraintumourcharity.org, call us on 01252 749043 or email fundraising@thebraintumourcharity.org

About this fact sheet

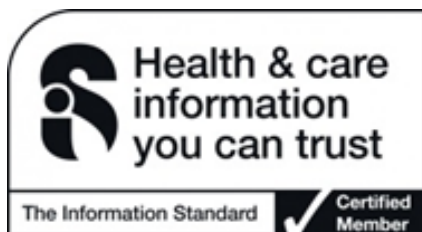


This fact sheet has been written and edited by The Brain Tumour Charity's Support and Information Team and is supported by the Children's Cancer and Leukaemia Group (CCLG).

The accuracy of medical information has been verified by 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



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