

# What is a brain tumour?

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 this process is disrupted, the cells can begin to grow in an uncontrolled way, creating a lump of cells called a tumour.

Brain and spinal tumours are central nervous system (CNS) tumours. They are usually named after the type of cell they start from and/or their location in the brain.

This fact sheet gives a brief outline of some of the most common types of brain tumour. If you have been diagnosed with a type of brain tumour not covered in this fact sheet, please contact our Information and Support team.

# In this fact sheet:

- ☐ What is a brain tumour?
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# What is a brain tumour?

A brain, or other CNS, tumour is an abnormal growth caused by cells dividing in an uncontrolled way.

There are over 130 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 the part 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.

Brain tumours are now generally referred to by health professionals according to their grade and their molecular/genetic make-up, rather than the terms you may have heard before - 'benign' and 'malignant'. This is because brain tumours may change grade over time.

# How are brain tumours graded?

Brain tumours are graded 1-4 according to their behaviour, such as the speed at which they are growing.

**Grade 1 and 2 tumours** are low grade, slower growing and less likely to spread to other parts of the brain.

There is also less chance of them returning if they can be completely removed by surgery or if the tumour cells are completely destroyed by other types of treatment, such as radiotherapy and chemotherapy.

They are sometimes still referred to as benign. The term benign is less used nowadays as it can be misleading. These low grade brain tumours can still be serious.

A tumour of any grade can cause harm by pressing on and damaging nearby areas of the brain, nerves and blood vessels. They can also block the flow of the cerebrospinal fluid (CSF) that nourishes and protects the brain, causing a build-up of pressure on the brain.

**Grade 3 and 4 tumours** are high grade and faster growing. They are more likely to spread to other parts of the brain (and rarely the spinal cord) and may come back, even if intensively treated. They can sometimes still be referred to as malignant.

They cannot usually be treated by surgery alone, but often require other treatments, such as radiotherapy and/or chemotherapy.

Some tumours contain a mixture of cells with different grades. The tumour is graded according to the highest grade of cell it contains, even if the majority of it is low grade.

# How common is a brain tumour?

Each year in the UK, approximately 11,000 people are diagnosed with a primary brain tumour. This represents fewer than 2 out of every 10,000 people in the UK. As such, brain tumours are classed as rare.

# Types of brain tumour

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.

They are then graded according to the way they look and behave under the microscope and the way the cells have mutated (changed). This may also contribute to their name.

Increasingly their genes are examined to give further information about how they are likely to behave and how they are likely to respond to certain treatments. This fact sheet outlines some of the most common types. The following is not an exhaustive list. If you would like information on any other type, please contact us.

## By cell type

## Glioma

A glioma is a tumour of the glial cells. Gliomas are the most common type of brain tumour.

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.

There are different types of glial cells - the main types being astrocytes, oligodendrocytes and ependymal cells. As a result, gliomas can also be divided into different types.

Types of glioma include:

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

The term glioma may be used to describe any of these the tumour types. It is, however, not very specific, so the term astrocytoma, for example, gives a more detailed description of the cell type the tumour originates from.

## **Astrocytoma**

Astrocytomas are the most common type of glioma in adults (and children), accounting for 34% of all brain tumours. An astrocytoma can be low grade or high grade.

Low grade astrocytomas are called pilocytic astrocytomas (grade 1) or diffuse astrocytomas (grade 2).

High grade astrocytomas are called anaplastic astrocytomas(grade 3) or a grade 4 astrocytoma is referred to as a glioblastoma.

Glioblastomas are the fastest growing brain tumour and the most common type of brain tumour in adults. (You may hear it called, less commonly, glioblastoma multiforme or GBM).

#### Oligodendroglioma

This type of glioma develops from the oligodendrocytes, which produce the fatty, protective covering (called myelin) of the nerve cells in the brain, This helps nerve signals travel along the nerves more quickly.

Oligodendrogliomas can be slow growing (i.e. grade 2), or they can be faster growing (when they are referred to as anaplastic oligodendroglioma (grade 3). They occur most often in the temporal or frontal lobes.

For more information, see *The human brain* webpage and fact sheet.

They account for around 3% of all brain tumours and are most likely to be diagnosed in adults.

#### **Ependymoma**

This type of tumour develops from ependymal cells, which are found in the fluid-filled areas of the brain. Ependymal cells repair damage to nerve tissue and produce the cerebrospinal fluid (CSF) that helps to cushion and protect the brain. They also have little hair-like structures on their surface, which help with the flow of CSF and chemical messengers throughout the brain.

Ependymomas can be high grade or low grade, but the appearance of their cells under the microscope (which is how the grading is established) does not always fit with their behaviour. So the grade may not tell you how likely it is to grow or spread.

They are less common - about 2% of all brain tumours. They occur more often in young adults (and children), where they can occur in any part of the brain. In older adults they usually occur in the lower part of the spinal cord.

As the ependymal cells are in fluid-filled areas of the brain, a tumour of these cells can spread within the central nervous system (brain and spinal cord) via the fluid, although this is not common.

#### Unspecified or mixed glioma

It is also possible to have what is known as a mixed or unspecified glioma. This is a tumour where either the doctor cannot tell what type of cell the tumour has, or where the tumour contains more than one type of glial cell (astrocytes, ependymal cells and oligogodendrocytes). An example of this is an oligoastrocytoma.

The treatment given is based on the highest grade of cell type within the tumour.

Mixed glioma account for around 6% of brain tumours.

However, the diagnosis of unspecified or mixed glioma is now less likely to happen, as we understand more about the molecular/genetic make-up of these tumours. In most cases, molecular testing can show if a previously named oligoastrocytoma can be classed as an astrocytoma or an oligodendroglioma.

# Vestibular Schwannomas

A schwannoma is a tumour that develops from the Schwann cells that cover the nerves and form the nerve sheath.

They can occur on any nerve in the body, but are commonly found on the nerve that controls hearing and balance and which runs from the ear to the brain.

These tumours are called a vestibular schwannoma or acoustic neuroma. Loss of hearing in one ear or problems with balance or dizziness are common symptoms of a vestibular schwannoma.

Schwannomas are found most often in older people. They tend to be slow growing, low grade tumours and do not generally spread from the site that they begin to grow from.

Rarely, they are associated with a genetic condition called neurofibromatosis type 2 (NF2), in which case they are usually diagnosed in much younger people.

# Embryonal tumours

Embryonal 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 part of the embryo that develops into the brain and spinal cord, is called the 'neuroectoderm', but some cells do not develop and specialise the way other cells do and so they appear "primitive".

Normally these primitive cells are harmless within the body, but rarely they can develop into tumours.

Embryonal tumours are more common in children and young adults. Medulloblastoma is the most common of these childhood embryonal tumours.

#### Medulloblastoma

Medulloblastomas originate from poorly developed embryonal (primitive) brain cells. The majority of them start in the cerebellum. This is a structure towards the back of the brain that controls balance and co-ordination).

Medulloblastomas are high grade (grade 4) and fast growing. They can spread to other parts of the brain through the cerebrospinal fluid (CSF).

They are the second most common of all brain tumour in children and the most common high grade tumour in children. They are also diagnosed in young adults.

## **Germ cell tumours**

Germ cell tumours grow from germ cells that form part of the embryo and develop into the reproductive system. They are also called embryonal tumours.

Most occur outside the brain, but those that do grow in the brain are most often in the area close to the pituitary gland and the pineal gland at the base of the brain.

These tumours are often in the middle of the brain and can block the flow of fluid (CSF) around the brain and spinal cord. This causes headaches and sickness and as a result these tumours are often picked up early when they are still small. They also sometimes produce chemicals that can be picked up on blood tests (markers) and this can also help to diagnose these tumours quickly.

They represent around 1 - 2% of all brain tumours. About half of these occur in children and young adults (10 - 20 years old).

# Haemangioblastomas

Haemangioblastomas are tumours that grow from blood vessel cells. They are low grade (grade 1), very slow growing and do not spread. However, they can grow in the brain stem and then they are very difficult to treat.

They represent about 2% of brain tumours.

People/families with a genetic condition called Von Hippel-Lindau Syndrome inherit a pre-disposition (proneness) to developing multiple haemangioblastomas. These can occur in the cerebellum, brain stem or spinal cord, as well as other areas, such as the kidneys

See *The Human brain* webpage and fact sheet for more information about these areas of the brain.

## By location in the brain

# Meningioma

A meningioma is a type of brain tumour that begins in the meninges. The meninges are a set of three membranes that cover and protect the brain and spinal cord. They occur most often in the membranes covering the cerebrum or cerebellum.

For more information, see *The human brain* webpage and fact sheet.

The majority of meningiomas are low grade.

However, some meningiomas can still cause some harm. If they are close to large blood vessels, have grown into the surrounding brain tissue or disrupted the flow of CSF around the brain, they will need to be treated and may be given a higher grade.

About one quarter (25%) of brain tumours in adults are meningiomas. They are more common in women and in older adults.

# Pituitary adenoma

A pituitary adenoma is a tumour that develops from the tissue of the pituitary gland. The pituitary is a gland that is found towards the base of the brain. It controls other glands within the body that, in turn, control many of the body's functions.

About 8% of tumours are in the pituitary gland. They are more common in older people and tend to be low grade, slow growing tumours that rarely spread.

Some pituitary adenomas can cause unusual effects by producing too much of one of the pituitary hormones. The pituitary gland sits very near to the optic nerves that control vision. Treatment will partly depend on whether your pituitary adenoma is causing problems with vision and if it is producing hormones or not.

# CNS lymphoma

A lymphoma is a tumour caused by the uncontrolled growth of the lymph cells (a type of white blood cell). Lymph cells help the body fight infection.

There are many different types of lymphoma - usually they form in the lymph nodes around the body.

Rarely, a lymphoma can form in other places, such as the central nervous system (CNS), is made up of the brain and spinal cord.

They can form either as a mass that presses on the brain or spinal cord from outside or, less commonly, as a mass or spread within the meninges (the membranes covering the brain).

These lymphomas are called a primary cerebral lymphoma or a primary CNS lymphoma.

They are usually high grade and are a type of non-Hodgkin lymphoma called diffuse large B cell non Hodgkin lymphoma. (Hodgkin lymphoma is a different illness entirely). It is important for your doctors to tell you which type of lymphoma you have.

CNS lymphomas represent just under 5% of brain and spinal cord tumours. They are usually treated differently to other brain tumours.

# 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 and can leave you feeling helpless and frustrated.

# Brain tumours are nobody's fault.

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.

#### 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 risk of developing a brain tumour:

- If an immediate family member (parent or sibling) has a tumour of the CNS you may have a slightly higher chance of developing a brain tumour, although the risk is still very low.
- If you have one of the following genetic conditions your risk of developing a brain tumour is increased:
  - neuro-fibromatosis (NF) types 1 or 2
  - Li Fraumeni syndrome
  - Von Hippel-Lindau syndrome
  - Turner syndrome

#### **Radiation**

We know that radiation does increase the risk of any tumour developing. Therefore, the risk of developing a meningioma or glioma is higher if you had radiotherapy to the head as a child, particularly before the age of five.

Due to the known risk of learning difficulties, health professionals try to avoid giving radiation to the brain of a young child under 3 years old.

Radiotherapy is not given without very careful consideration and you would not have been given radiotherapy unless the expected benefits outweighed any potential risks to you.

# How are brain tumours treated?

Several factors influence the decision as to which treatment will best help you.

A team of specialised health professionals will consider your individual diagnosis and take account of factors, such as the size and location of the tumour, the type of tumour you have and how quickly

it is growing. They will also consider your age and general health. You may hear this team being referred to as the Multi-Disciplinary Team or MDT.

# For more information, see our MDT webpage and fact sheet.

**Surgery** will often be used to remove as much of the tumour as possible. Surgery may be performed if the location of the tumour is causing problems, such as disrupting the flow of CSF, causing the brain to swell (cerebral oedema) or compressing the nerves.

As there is not much room in the skull for anything extra, the presence of a brain tumour can cause the pressure within the skull to increase. 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 you may not need surgery straight away or even at all.

For more information, see our *Neurosurgery* and *Watch* and wait (active monitoring) webpages and fact sheets.

Other treatments, such as **radiotherapy** and **chemotherapy** may be used - on their own, in combination or after surgery to try to remove any remaining tumour cells.

Forms of radiotherapy are also considered if a person has many tumours, such as in Von Hippel-Landau Syndrome, described earlier in this fact sheet.

For more information, including information about duration of treatment, please see our *Radiotherapy* and *Chemotherapy* webpages and fact sheets.

Specialist radiotherapy, such as Cyber knife<sup>®</sup> may be used to treat a small number of brain tumours that cannot be reached by surgery.

For more information, see our *Stereotactic radiotherapy* webpage and fact sheet.

A further type of treatment, that a few people may be able to have, is **Proton Beam Therapy** (PBT).

PBT is a type of radiotherapy that uses a beam of protons ('positively charged subatomic particles ') to target the tumour in a very precise way, reducing the harm to healthy tissue.

However, PBT is only suitable for a few types of brain tumour. It works best for smaller tumours and those where the edges are clearly defined.

There is also some debate about whether it is any better than conventional well-targeted radiotherapy. It is not currently available in the UK for use in brain tumours, though it can be received outside the UK through the NHS where appropriate and in certain circumstances.

The Department of Health has announced two NHS 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. There are also some private facilities being developed.

Given the complex nature of the treatment and facilities, PBT won't be fully available in England until 2018 onwards. 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 you do not meet this criteria you will not be referred on the NHS.

For further information, see the *Proton Beam Therapy* webpage and fact sheet.

It is important to know that your medical team will tailor your treatment to provide the treatment that is the best for you. This could mean that you meet other patients who have the same tumour but who are receiving different treatments. The reasons for this could be due to several factors.

If you are worried or concerned at all, you should speak to your medical team who will be able to help you understand the treatment decisions that have been made. If you feel you are not getting the correct treatment you can ask for a second opinion.

More information about this can be found on the *Getting a second* opinion webpage on our website.

# What effects can I expect to experience from a brain tumour?

This is difficult to say. With over 130 types of brain tumour, the effects anyone experiences will depend to a large extent on the type, grade and location of your tumour. Some of the effects will be a result of the brain tumour itself, whilst others will be due to the treatment you receive and when you receive it.

In addition, every person will react differently depending on their general health, age and genetic make-up.

However, for information about what effects you may experience, please see our webpages/fact sheets on: *Symptoms of a brain tumour in adults*; *Neurosurgery*; *Chemotherapy*; *Radiotherapy*; and *Steroids*.

# It is important to remember

Although brain tumours can be difficult to spot as their symptoms can be very similar at first to many common ailments, they are very rare.

Please see our *Symptoms of a brain tumour in adults* webpage/fact sheet; and for young adults and children, see our HeadSmart website: www.headsmart.org.uk

# What if I have further questions or need other support?

You can contact our Information and Support Team in the following ways:



0808 800 0004

(Free from landlines and most mobiles: 3, O2, EE, Virgin and Vodafone)



support@thebraintumourcharity.org



#### Live Chat

Get in touch with us online via thebraintumourcharity.org/live-chat



Join one (or more) of our closed Facebook groups: bit.ly/FBSupportGroups



thebraintumourcharity.org/getsupport

Want to make a difference through your diagnosis? BRIAN can help. Sign up at: thebraintumourcharity.org/BRIAN

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# About this information resource

The Brain Tumour Charity is proud to have been certified as a provider of high quality health and social care information by The Information Standard - an NHS standard that allows the public to identify reliable and trustworthy sources of information.

Written and edited by our Information and Support Team, the accuracy of medical information in this resource has been verified by leading health professionals specialising in neuro-oncology.

Our information resources have been produced with the assistance of patient and carer representatives and up-to-date, reliable sources of evidence.

We hope that this information will complement the medical advice you've already been given. Please do continue to talk to your medical team if you're worried about any medical issues.

If you'd like a list of references for any of our information resources, or would like more information about how we produce them, please contact us.

We welcome your comments on this information resource, so we can improve. Please give us your feedback via our Information and Support Team on 0808 800 0004 or support@thebraintumourcharity.org

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# **About The Brain Tumour Charity**

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 worldwide, raise awareness of the symptoms and effects of brain tumours and provide support for everyone affected to improve quality of life.

We wouldn't be able to make the progress we have without the incredible input we receive from you, our community.

Whether it's reviewing our information resources, campaigning for change, reviewing research proposals or attending cheque presentations, everything you do helps to make a difference.

To find out more about the different ways you can get involved, please visit thebraintumourcharity.org/volunteering

# We rely 100% on charitable donations to fund our work.

If you would like to make a donation, or find out more about other ways to support us, including leaving a gift in your Will or fundraising through an event, please get in touch:



thebraintumourcharity.org/get-involved call us on 01252 749043 or email fundraising@thebraintumourcharity.org









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