The BRAIN TUMOUR CHARITY

Research | Awareness | Support

Astrocytoma

Astrocytomas are tumours that grow from a type of cell in the brain called an astrocyte. They're the most common type of a group of brain tumours called gliomas and account for about one third of all brain tumours.

There are various types of astrocytomas, each of which affects both adults and children.

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

This fact sheet doesn't deal in detail with grade 4 astrocytoma (glioblastoma/GBM) as this is explained in a separate fact sheet.

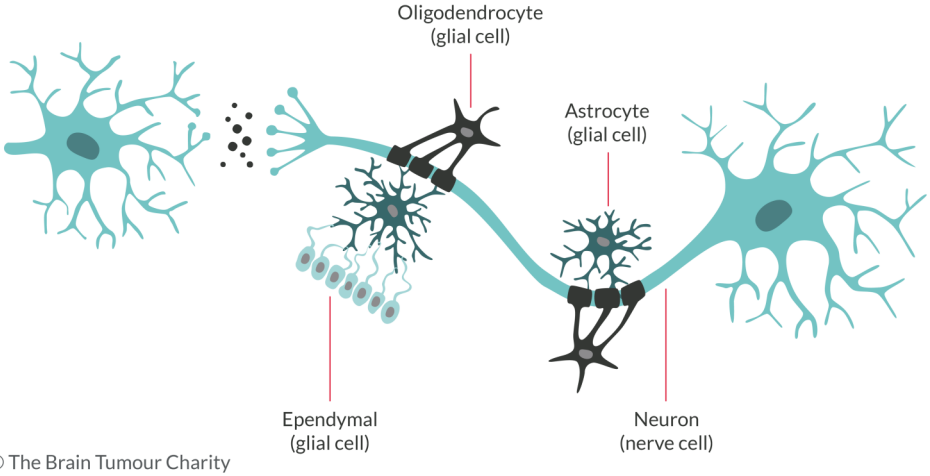
In this fact sheet:

What is an astrocytoma?

Astrocytomas are tumours that grow from a type of cell in the brain called an astrocyte.

Throughout the brain and spinal cord we all have nerve cells called neurons, which transmit messages (electrical and chemical signals) to, from and within the brain. Surrounding the neurons are cells called glial cells, that support and protect the neurons by providing them with oxygen and nutrients and removing dead cells.

An astrocyte is a type of glial cell. There are three main types of glial cells - astrocytes, oligodendrocytes and ependymal cells.



Astrocytes are the most common type of cells in the brain. As well as supporting and protecting neurons, they also help to pass messages between the neurons and are therefore vital in processing information in the brain.

Like all cells, astrocytes normally grow in an orderly, controlled manner. However, if this process is upset or disrupted for some reason, they can continue to grow and divide when they shouldn't, causing a lump (a tumour) to form. (Tumours are an abnormal growth caused by cells dividing in an uncontrolled way.)

Brain tumours are usually named according to the type of cell they grow from and/or the part of the brain they grow in. So when astrocytes form a tumour it's known as an astrocytoma.

You may also hear it called a type of glioma. Tumours that grow from any glial cell are collectively called gliomas. A tumour that grows from the astrocytes is therefore a glioma, but is more accurately described as an astrocytoma.

Types of astrocytoma

Brain tumours are graded by the World Health Organisation (WHO) from 1-4, according to how they behave, i.e. how fast they grow and how likely they are to spread within the brain.

Tumours graded 1 and 2 are slow-growing, and are sometimes called benign or low grade. This term is used less often nowadays as it's not thought to be helpful in describing the tumour, as these low grade tumours are still serious.

Tumours graded 3 and 4 are fast-growing, more aggressive tumours, sometimes called malignant or cancerous, meaning they're more likely to get bigger more quickly and sometimes spread to other parts of the brain or spinal cord.

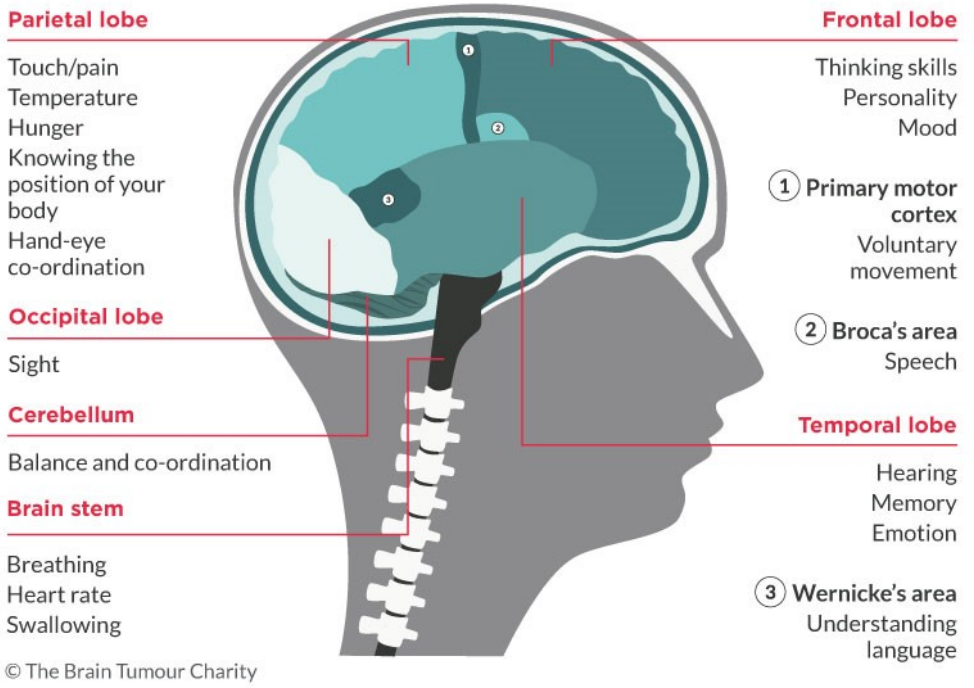
Astrocytomas can be any grade, from 1-4.

Grade 1 astrocytoma (pilocytic astrocytoma)

Grade 1 astrocytomas are called pilocytic astrocytomas. (Pilocytic means the cells are elongated and look hair-like.) These are slow growing, relatively contained and unlikely to spread to other parts of the brain. They're also unlikely to return if they're completely removed by surgery.

They're most often found in children and young adults under the age of 20 and are equally common in males and females. They're very rare in adults over 50 years.

They tend to occur in the cerebellum - the part of the brain at the back that controls balance, but they can also occur in the optic pathways. These are the pathways from the eyes to the visual cortex in the occipital lobe at the back of the brain, which is responsible for sight.



Lobes of the brain and their functions

Pilocytic astrocytomas also occur in around 10% of people who have neurofibromatosis type 1 (NF1). This is a genetic condition you're born with that causes tumours to grow along your nerves.

Grade 2 astrocytoma (diffuse astrocytoma)

The most common grade 2 astrocytoma is called a diffuse astrocytoma. (Diffuse means it doesn't have well-defined edges.) These are slow-growing, but they can come back as a higher, grade 3 anaplastic astrocytoma or a grade 4 astrocytoma (glioblastoma).

They occur most often in adults between the ages of 20 and 45. They're more common in males than females.

Grade 3 astrocytoma (anaplastic astrocytoma)

A grade 3 astrocytoma is called an anaplastic astrocytoma. (Anaplastic means the cells divide rapidly and don't resemble normal cells in structure or function.) They're fast-growing and often called malignant or cancerous. They often recur following initial treatment in a more advanced form, i.e. grade 4 astrocytoma (glioblastoma).

They're more common in adults between the ages of 30 and 70 and are more common in males.

Grade 4 astrocytoma (glioblastoma)

Grade 4 astrocytomas are usually called glioblastoma. You may sometimes hear them called glioblastoma multiforme, or GBM for short, though these terms are less used nowadays.

See the separate *Glioblastoma* webpage and fact sheet for more information.

What causes astrocytomas?

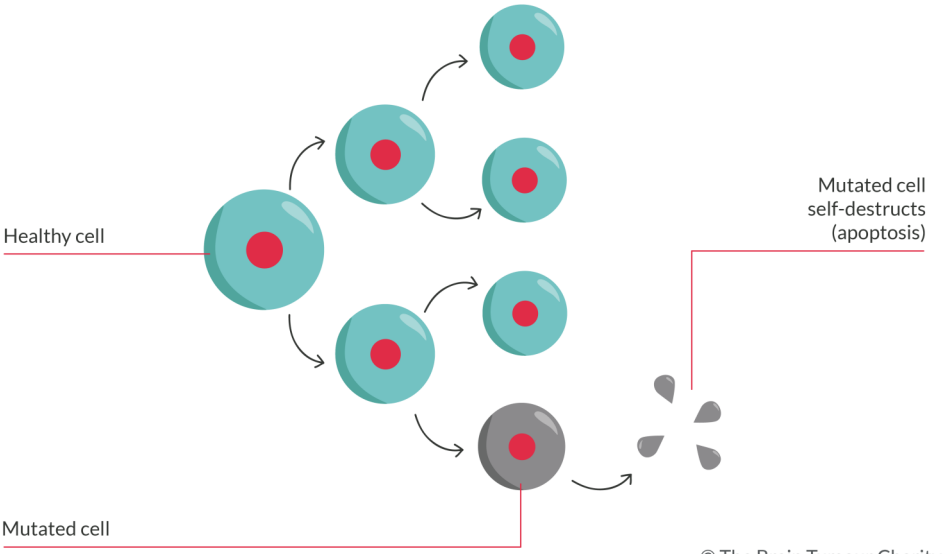
The first thing to mention is that there's nothing you could have done, or avoided doing, that would have prevented you from developing an astrocytoma.

As with most brain tumours, the cause is not known. However, much research is being carried out into possible causes, focussing on our genes.

Genes

Astrocytomas, like all brain tumours, are the result of uncontrolled growth of brain cells.

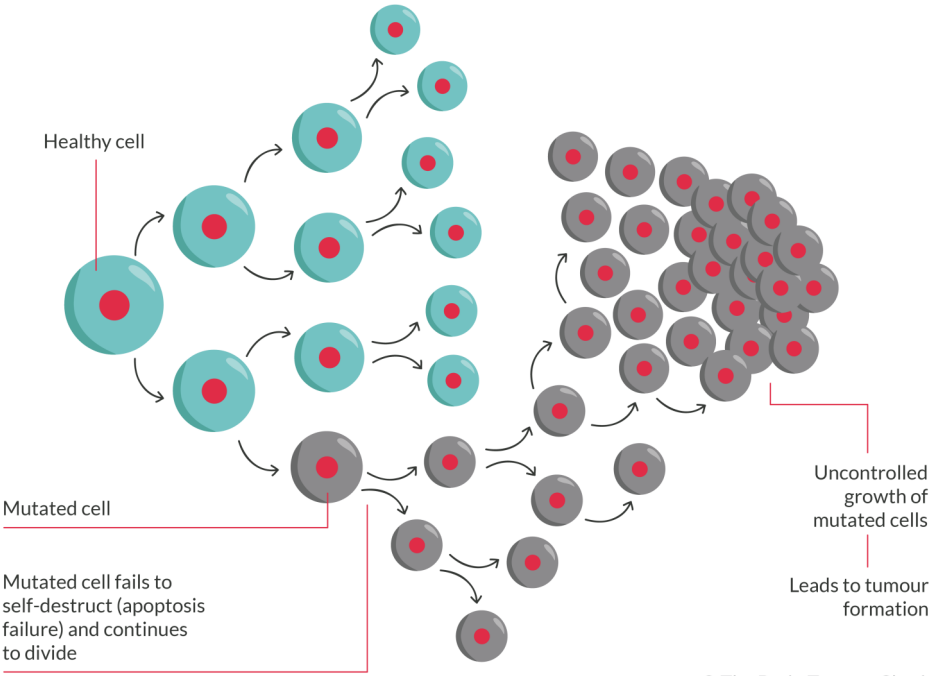
Normal cells grow, divide and die in a controlled way, in response to signals from your genes, which are present in all your cells. These signals travel down a particular 'pathway' to tell the cell when to grow and when to stop growing. Even if these signals are missing or are in the wrong combination, our bodies also have other ways (checkpoints) to stop cells dividing in an uncontrolled way.



Normal cell division

Mutations (changes) in specific genes in the DNA (genetic material) of a cell can lead to tumour growth by altering the pathway and making the cell behave as if it's receiving a growth signal, even if it's not.

Alternatively, the mutations can inactivate the checkpoints that would normally stop the cell from dividing. As a result, any cells affected will continue to divide and can develop into a tumour.



Altered cell division leading to tumour formation

Different grades of astrocytoma have mutations in different genes and research funded by The Brain Tumour Charity has played a large part in identifying some of these.

These include:

- Grade 1 pilocytic astrocytomas often have a mutation in genes called NF1 and BRAF.
- A mutation in the IDH-1 gene has also been found in a large number of astrocytomas (as well as other types of glioma) across all grades. Those that have the mutation tend to have a better prognosis than those that don't.
- Other genetic changes which may be important in astrocytomas are mutations in the ATRX gene and loss of some genes from particular chromosomes, known as 1p/19q co-deletion.
- Grade 2 diffuse astrocytomas and grade 3 anaplastic astrocytomas often have mutations in genes called TP53 and PDGFR.

It's hoped that this information will help in the development of more personalised treatments.

See *Biomarkers and future treatments* section later in this fact sheet.

Cells

Grade 3 anaplastic astrocytomas often develop from grade 2 diffuse astrocytomas that have become more advanced and malignant.

However, it's not clear whether this is the case for all or just most anaplastic astrocytomas, as there are a number of cases of grade 3 anaplastic astrocytomas that have grown from new. You may hear this called de novo.

How is an astrocytoma diagnosed?

If your doctor (GP or A&E doctor) suspects you have a brain tumour, they'll refer you to a specialist - a neurologist or neurosurgeon (specialists in brain and nerve disorders). If it's your child that they suspect has a tumour, they'll refer them to a paediatrician.

Neurological examination

The specialist will ask questions about your health and give you a physical examination. They'll also test your nervous system (called a neurological examination). This involves looking at your vision, hearing, alertness, muscle strength, co-ordination, and reflexes.

They may also look at the back of your eyes to see if there's 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.

Scans

You'll then have one or more further tests, such as an MRI (magnetic resonance imaging) or CT (computerised tomography) scan to establish whether a brain tumour is present.

For information about these scans, please see the *Scans* webpages and fact sheet.

For some people, their first symptom may be a seizure, so they're seen as an emergency. In this case they may be given a scan as their first test, after which their case will be referred to a neuro-oncology MDT (multi-disciplinary team) followed by a consultation with the neurologist/neurosurgeon.

See the *Multi-Disciplinary Team (MDT)* webpage and fact sheet for more information.

Some GPs can refer you for a scan directly, or you may have been admitted to hospital with a problem and a brain scan was arranged to investigate this further.

If, following the scan, a tumour is found, you may be given a body scan to establish if your tumour is a primary brain tumour (one which started in the brain) or a secondary tumour and there's a primary tumour elsewhere in the body.

Surgery

Where possible, the whole tumour, or as much as is safely possible, will be surgically removed. This is called a resection of the tumour and is done during an operation called a craniotomy.

If only part of the tumour can be safely removed, this is called debulking or maximal safe resection.

See the *Neurosurgery for adults with brain tumours* webpage and fact sheet for more information.

Biopsy

If the tumour is a primary tumour and is in an area of the brain which can be operated on, a biopsy (small sample of the tumour) may be taken from your tumour to aid diagnosis of the tumour type.

The neurosurgeon will always take a biopsy as part of a craniotomy, but if the tumour cannot be removed, they may take a biopsy through a small hole in the skull.

It's important to realise that a biopsy is an operation that takes several hours. The risks will be explained to you by your surgical team.

Biobanking

In both cases of biopsy or surgery, you may like to ask**, before your operation,** about the possibility of biobanking some of the tissue from your tumour.

This means storing some of your tumour tissue, so you can be a candidate for clinical trials in the future and have any relevant genetic (biomarker) tests.

It can also be used for research to help find better treatments and ultimately a cure.

Some trials and treatments may require the tissue to be stored in a certain way, i.e. by flash freezing, rather than by storing in paraffin wax as is common. Make sure you mention this when asking about biobanking.

Speak to us and to your healthcare team if this is something you're interested in doing. The Brain Tumour Charity Information and Support Line:0808 800 0004 or ***support@thebraintumourcharity.org*** Or get in touch online via: ***thebraintumourcharity.org/live-chat***

Laboratory analysis

Following biopsy or surgery, cells from the tumour will be analysed in a laboratory by a neuropathologist.

See the *Multi-Disciplinary Team (MDT)* webpage and fact sheet for more information.

The neuropathologist will examine the cells, looking for particular cell patterns that are characteristic of the different grades of astrocytoma.

Confirming the diagnosis of the different grades of astrocytoma can be difficult as low grade and high grade astrocytoma cells can look very similar. However, detection of the various gene mutations, previously mentioned in this fact sheet, are now being used to aid:

- accurate diagnosis
  e.g. BRAF fusion gene - used for diagnosing pilocytic astrocytomas
- prognosis
  e.g. IDH1 gene - used to give information about possible length of survival.

Accurate diagnosis is important as it allows your medical team to give you information about how the tumour could behave in the future, and also to recommend treatment options. This could include a clinical trial.

Note: Unfortunately there are not enough trials running for people with brain tumours, particularly low grade tumours. The Brain Tumour Charity is committed to improving this and making sure every patient has the opportunity to be involved in research.

Biomarker testing

As part of this analysis, you may like to ask about biomarker testing. This is where the doctors look for markers (changes) in certain genes in the tumour cells that may indicate how well you'll respond to certain treatments.

See the *Biomarker tests and future treatment* section later in this fact sheet.

How are astrocytomas treated?

The treatment for astrocytomas depends on the grade of the tumour, as well as its size and location.

Grade 1 pilocytic astrocytoma

Surgery - complete tumour removal

Where possible, treatment for grade 1 pilocytic astrocytomas in adults is normally surgery. The aim is to remove as much of the tumour as possible. How much can be removed will depend on where the tumour is in the brain.

If the tumour is in the cerebrum or cerebellum they can often be removed completely. You may hear this called complete or total resection.

See previous diagram on page 4 or *The human brain* webpage and fact sheet .

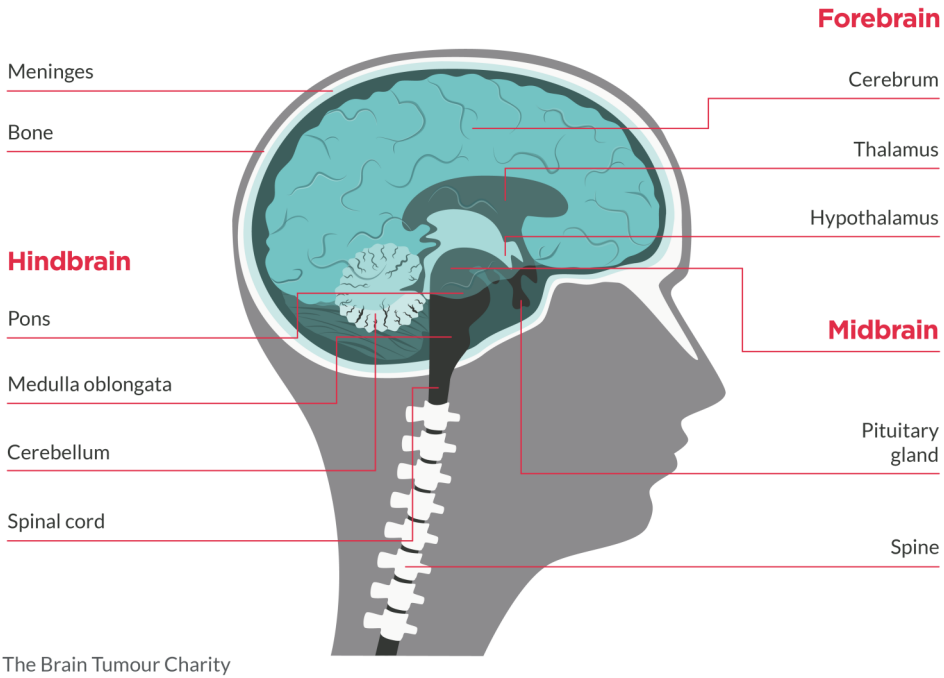
If the tumour is completely removed, more treatment may not be needed. Your hospital will continue to monitor you with regular MRI scans, but no treatment will be given, unless you develop symptoms, any symptoms you have worsen or your scan changes. This is often called watch and wait or active monitoring.

See the *Scans (adults)*, *Scans for children* and the *Watch and wait (active monitoring)* webpages and fact sheets for more information.

With tumour locations other than the cerebrum or cerebellum, complete removal may still be possible, but the benefits need to be weighed up against various possible after-effects.

For example, if the tumour is in the optic nerve, complete removal may be achieved, but it can result in blindness in the affected eye.

Or, if the tumour is in one of the midline structures (for example, the hypothalamus, brain stem or spinal cord) surgery can affect the functions these areas control, particularly in children younger than 2 years old. These functions include breathing, sleep and body temperature.



Main parts of the brain.
The brain stem includes the pons, medulla oblongata and mid brain.

Surgery - partial tumour removal

In some cases, complete removal of the tumour may not be possible. It may be located in a difficult area of the brain to operate on, or near important parts of the brain where surgery could do more harm than good. In this case, the surgeon will remove as much of the tumour as they can. This is known as debulking or partial resection.

Depending on how much of the tumour is removed, you may be put on a watch and wait or active monitoring approach after surgery.

See the sections further on in this fact sheet and also the *Watch and wait (active monitoring)* webpage and fact sheet.

Alternatively you may be given radiotherapy after your surgery, though doctors try to avoid this in people with the genetic condition NF1 and also in children under 3 years old. Occasionally chemotherapy may also be given.

See the *Chemotherapy*, *Radiotherapy*, *Chemotherapy for children*, or *Radiotherapy for children* webpages and fact sheets.

Further surgery may be needed later to remove the rest of the tumour that was left, or if it starts to regrow. You may also need further surgery if the tumour is blocking the flow of the cerebro-spinal fluid (a condition called 'hydrocephalus'), to insert a shunt to remove the excess fluid from the brain.

Chemotherapy

For children, chemotherapy is often the first line of treatment.

See the *Chemotherapy for children* webpage and fact sheet.

Watch and wait (active monitoring)

In some cases, your medical team may decide not to use surgery initially, but to use the watch and wait approach. This is more accurately now called active monitoring, and may also be used after surgery.

This may be used if the tumour:

- is growing very slowly
- is not causing any symptoms
- is causing only a few symptoms that are well controlled or which are not badly affecting your quality of life.

The reason for adopting this approach may be that it's felt that surgery carries more risks than giving no initial treatment. This may feel like they're 'doing nothing'. However, they'll monitor your tumour and, where necessary, treat any symptoms.

In this situation, treatment may not be needed for many months or even years. Some people may never need any further treatment.

Some tumours have also been known to reduce in size with no apparent cause. This is known as spontaneous regression.

If your symptoms worsen, or the tumour changes or starts to grow, your medical team will then look at other treatment options, such as surgery.

Grade 2 diffuse astrocytoma

As with pilocytic astrocytomas, surgery is usually the first treatment for diffuse astrocytomas in adults. If they can be completely removed or almost completely removed, then treatment is the same as pilocytic astrocytomas.

However, as these tumours are more diffuse (the edges are less well-defined), complete removal is often not possible and more of the tumour is often left behind. These tumours are more likely to regrow.

Surgery, therefore, is more often followed up with radiotherapy and chemotherapy. This is particularly the case if you have symptoms such as weakness in an area of your body.

For children, particularly young children, chemotherapy, rather than surgery, is usually the first treatment given.

Grade 3 anaplastic astrocytoma

As these tumours are faster growing and more aggressive, the usual course of treatment is surgery followed by radiotherapy and sometimes chemotherapy.

If you're relatively fit, a long course of radiotherapy over several weeks may be suggested - if not, a shorter course may be more suitable. Radiotherapy is very rarely used in children under 3 years.

Your oncologist may also recommend a drug called temozolomide (Temodal®). Temozolomide stops the tumour cells copying their DNA (genes), which needs to happen before the cells can divide. This in turn stops the tumour cells dividing.

If a high grade astrocytoma is suspected on an MRI scan before your operation, your healthcare team may discuss the option of inserting chemotherapy wafers (Gliadel®).

These are polymer wafers, coated in a chemotherapy drug (usually carmustine) and inserted into the brain during surgery. The polymer gradually dissolves over 2-3 weeks, releasing the chemotherapy drug directly into the brain. They are used to target cells which are beyond the region removed by surgery.

However, they're only licensed for use in adults with high grade gliomas (such as anaplastic astrocytomas) and recurrent glioblastoma (GBM), so their use in children is extremely rare.

NICE guidelines also mean that these can only be used when the surgeon is confident that at least 90% of the tumour has been removed and that the fluid-filled spaces in the brain (ventricles) have not been affected by the tumour. (NICE is the National Institute for Health and Care Excellence.)

In addition, some neurosurgeons don't regularly use the wafers, as they're concerned about possible increased wound infection rates after surgery.

For more information, see the *Chemotherapy* and the *Chemotherapy for children* webpages and fact sheets.

Grade 4 astrocytoma (glioblastoma)

Information about this type of astrocytoma can be found in a separate fact sheet.

See the separate *Glioblastoma* webpage and fact sheet for more information.

Biomarker tests and future treatment

A biomarker is a biological marker or indicator, such as a change in a gene in a tumour's DNA, that can indicate how likely the tumour is to respond to treatment or how likely it is to progress.

See the *Biomarkers* webpage and fact sheet for more information.

MGMT methylation test

This test involves looking at how much of a particular protein involved in DNA repair (MGMT) is present in the tumour cells.

MGMT can disrupt the action of some chemotherapy drugs, such as temozolomide, so high levels of MGMT means that chemotherapy is likely to be less effective. This information helps to plan suitable, individualised treatment.

The test is only suitable for certain types of tumour, of which anaplastic astrocytoma is one.

The MGMT methylation test is now routinely carried out in many neuro-oncology centres.

IDH-1 test

This test looks for a mutation in a gene known as IDH-1.

For people with high grade types of certain gliomas, including astrocytomas, mutations of the IDH-1 gene are often linked with longer-term survival rates.

It's not as clear how mutations of the IDH-1 gene link to outcomes for people with low grade brain tumours, but grade 2 astrocytomas that don't carry the mutation are likely to behave more aggressively.

In addition to long-term survival rates, scientists have looked at whether the IDH-1 gene mutation predicts treatment outcomes. However, while this research is ongoing, there's no difference in treatment based on your IDH status.

If you're interested in IDH-1 testing, please speak to your neuro-oncologist for information and advice.

Future treatment

As we gain greater understanding of the role played by genes and signalling pathways within cells in the development of astrocytomas (*see previous sections of this fact sheet*), possible drug treatments which target these processes may be developed that don't affect surrounding healthy tissue.

For example, work funded by The Brain Tumour Charity on the BRAF fusion gene and its pathway called MAP kinase, has led directly to clinical trials with drugs that target this pathway, in inoperable and recurrent pilocytic astrocytoma.

These drugs were chosen to be investigated as it follows the successful use of similar drugs to treat malignant melanoma (a type of skin cancer), which also has defects in the MAP kinase pathway.

For more information, see our *Emerging treatments* webpage.

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:
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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***

Your notes:

Your notes:

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:

Visit
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call us on 01252 749043 or email
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