

Brain and other central nervous system tumours - fact sheet



The central nervous system (CNS) is the part of the nervous system that consists of the brain and spinal cord.

There are many different types of tumours that can arise in the CNS. They are named according to the type of cell or part of the brain/spinal cord from which they arise.

Tumours which start in the brain or spinal cord are called primary tumours (or cancers). Other types of cancers which spread to the brain from other parts of the body are referred to as secondary tumours (or metastases). This section focuses only on primary brain and spinal cord tumours.

Brain and other CNS tumours occur when abnormal cells in these organs grow in an uncontrolled way. As they grow and spread these tumours cause a wide range of symptoms depending on which parts of the brain or spinal cord are affected.

There are many different types of cells in the brain and other parts of the CNS, and each of these can develop into different types of tumours that need to be treated in different ways. The most common types of brain and other CNS tumours in children are:

- **Gliomas** – these develop from glial cells. There are several types of glial cells which give rise to different types of gliomas:
 - **Astrocytomas** develop from a glial cell called an astrocyte. This is a cell that supports the nerve cells in the brain. The terms 'astrocytoma' and 'glioma' are often used interchangeably. Astrocytomas/Gliomas are categorised as low or high grade according to how 'aggressive' the tumour cells look under a microscope. The Grade I and II are low grade tumours whereas Grade III and IV are considered high grade. The most common types of astrocytomas are:
 - **Juvenile Pilocytic Astrocytoma (JPA)** is the commonest low grade tumour in children. It usually occurs in cerebellum or the backpart of the brain.
 - The commonest high grade tumours are **Glioblastoma Multiforme (GBM)** which is a grade IV tumour, and Anaplastic Astrocytoma (Grade III) are both high grade tumours. These generally develop in the upper part of the brain.
 - **Diffuse Intrinsic Pontine Gliomas (DIPGs)** develop in the brain stem. This is the part of the CNS that “connects” and lies between the brain and the spinal cord. It acts as the control centre for vital body functions such as the heart rate and breathing.
 - **Optic pathway gliomas** arise either in the optic nerve, which connects the brain with the eyes or the vision pathways in the brain.
- **Ependymomas** develop from ependymal cells. These cells line the fluid-filled areas of the brain (called the ventricles) and spinal cord, through which the cerebrospinal fluid flows. Ependymomas are given different names, depending on where they occur in the brain. They can spread to other parts of the CNS.
- **Medulloblastomas** – these tumours develop from a type of nerve cell in the cerebellum (the area at the back of the brain that controls movement and coordination). They tend to be aggressive tumours and often spread to other parts of the brain or spinal cord.
- There are many **other types** of brain and spinal cord tumours. These are much rarer. They include atypical teratoid rhabdoid tumours (ATRT), primitive neuroectodermal tumours (CNS-PNET), pineoblastoma and CNS germ cell tumours .





Risk factors

A risk factor is anything that increases a person's chance of developing a certain condition or disease, such as cancer. In adults, lifestyle and environmental factors (such as smoking or exposure to certain chemicals) can be significant risk factors for developing certain types of cancer. In children, very few risk factors have been identified that increase the chance of developing cancer. For most children with cancer, the underlying cause is unknown.

Even if your child has a risk factor, it does not mean they will develop cancer. Many children with a risk factor will never develop cancer, while others with cancer may have had no known risk factors. Even if a child with a risk factor develops cancer, it is usually hard to know how much that risk factor contributed to the development of their disease.

The causes of brain and other CNS tumours are not well understood, but factors associated with a higher chance of developing brain and other CNS tumours include the following.

Family history

In rare cases, an increased chance of developing brain and other CNS tumours runs in families. This is more likely to be the case if other people in your family had brain or other CNS tumours in their childhood. When these tumours run in families, the tumour usually develops at a younger age than in children with no family history of the disease.

Knowledge of your family's history relating to cancer may change over time.

Genetic conditions

Some types of genetic conditions can increase a child's chance of developing brain and other CNS tumours. In these cases, specific changes in one or several genes have been identified that contribute to cancer. These conditions can run in families, or they can start in the child from a fault in their DNA.

Conditions that are associated with childhood brain and other CNS tumours include:

- neurofibromatosis type 1 (von Recklinghausen disease)
- neurofibromatosis type 2
- tuberous sclerosis
- von Hippel–Lindau syndrome
- Li–Fraumeni syndrome
- Gorlin syndrome





- Turcot syndrome.

If your child is diagnosed with one of these genetic conditions, they will need specific follow-up. The health care team will advise which ongoing tests your child will need.

Cancers in children that are linked to genetic conditions may also affect the risk for other family members. Speak to your child's treatment team to see whether genetic counselling is recommended for you or your family.

For more information about genetic conditions, see the [children's cancer glossary](#) or the [Centre for Genetics Education](#).

Exposure to radiation

Children who have had radiation exposure to the head (usually to treat other cancers such as leukaemia) have an increased chance of developing brain tumours later in life.

Diagnostic imaging tests such as computed tomography (CT) scans and X-rays expose a person to a very low dose of radiation. The potential risk to an unborn baby or child from this type of radiation exposure is not known, so doctors usually avoid these tests for pregnant women or young children unless they are essential.

Symptoms

Symptoms of brain and other CNS tumours can depend on where the tumour is, how fast it is growing and the child's age. Some symptoms occur because the tumour creates pressure in the brain. Pressure in different parts of the brain can produce different symptoms.

Symptoms are not the same in every child, but can include:

- headaches
- nausea or vomiting
- problems with vision, hearing or speech
- problems with balance or coordination
- weakness or numbness in a part of the body, especially only on one side
- back pain
- changes in behaviour
- weight loss or weight gain for no reason





- seizures (fits)
- drowsiness or coma.

Very young children or babies may not be able to tell you their symptoms. Symptoms in young children can also include:

- irritability
- loss of appetite
- delays in development
- failure to thrive – where a child fails to gain weight appropriately
- decrease in physical or intellectual abilities, or losing abilities that they had before
- increase in head size or swelling of the soft spots of the skull.

Note about symptoms

Many conditions – including common childhood infections – can cause these symptoms, not just brain and other CNS tumours. If your child has any of these symptoms and you are concerned, talk to your child's doctor.

Diagnosis

Your child will have a number of tests to investigate their symptoms and confirm a diagnosis of a brain or other CNS tumour, including:

- medical history and physical examination, including a neurological examination
- blood tests
- urine tests
- medical imaging, which may include:
 - X-ray
 - computed tomography (CT) scan
 - magnetic resonance imaging (MRI)
 - positron emission tomography (PET) scan
- biopsy – where a small sample of the tumour is removed to be examined under a microscope
- lumbar puncture (spinal tap) – where a sample of cerebrospinal fluid is taken to be examined under a microscope.

These tests are explained in more detail in [How is cancer diagnosed?](#)





Staging

If your child is diagnosed with a brain or other CNS tumour, some of the diagnostic tests will also help to stage the tumour. Staging determines where the tumour is, how big it is and whether it has spread to other parts of the body. This is important to determine the outlook (prognosis) for your child, and to decide on the best options for treatment.

There is no standard staging system for childhood brain and other CNS tumours. Instead, tumours are staged based on a range of factors (such as size and location of the tumour) that are classified into different risk groups.

Treatment

Treatment and care of children with cancer is usually provided by a team of health professionals called a multidisciplinary team. Members of this team are specialists in children's cancers – they understand the differences between children's cancer and adult cancer, and each team member brings different skills in managing care to meet the needs of both you and your child.

The team will be led by a childhood cancer specialist (paediatric oncologist). Other members of the team depend on the age of your child and their type of disease, and may change over time as your child's needs change. A list of team members who might make up the multidisciplinary team can be found in [The treatment team](#).

Treatment for brain and other CNS tumours depends on the age of your child, the stage of the disease, the biological features of the cancer and other factors identified during diagnosis. Treatment will be tailored to your child's particular situation, and may involve one or more of the following (see [How is cancer treated](#) for more detail).

Surgery

Most children with brain or other CNS tumours have surgery to remove all or part of the tumour. Brain surgery can affect the normal functioning of your child's brain or nerves. However, the neurosurgeons who perform this surgery are extremely careful to avoid damaging your child's normal brain tissue.

If the entire tumour can be removed and it has not spread to other parts of the body, your child may not need any other treatment.

If the tumour has spread to surrounding areas, or is in a part of the brain or CNS that makes complete





removal of the tumour very difficult, surgery can still be used to remove as much of the tumour as is safely possible. This reduces the size of the tumour that needs to be treated with chemotherapy or radiation therapy, and may make these treatments more successful.

Reducing the size of the tumour through surgery can reduce pressure on the brain, which can help to relieve symptoms such as headaches, nausea, seizures and blurred vision.

Surgery can also be used to drain excess fluid to relieve pressure on the brain, or to insert a device that allows chemotherapy to be delivered directly into the cerebrospinal fluid (known as a ventricular access catheter).

After surgery, your child may have one or more tubes coming out of the surgical site to drain excess fluid out of the skull. These tubes are usually removed after a few days.

Chemotherapy

Chemotherapy uses anti-cancer medicines to destroy cancer cells. It is often given as a combination of medicines to try to prevent the cancer cells from becoming resistant to just one or two medicines.

Chemotherapy medicines are given together in courses, often over a few days. Once the body has recovered from the side effects, the next course is given. Most children receive multiple courses of chemotherapy .

Chemotherapy may be used for brain and other CNS tumours:

- after surgery (to destroy any remaining cancer cells)
- as the main treatment (if surgery is not a good option, or to avoid or delay using radiation therapy, or along with radiation therapy)
- in high doses with stem cell support (i.e. high-dose chemotherapy followed by a stem cell transplant).

Chemotherapy for many types of cancer is usually given intravenously (IV) or by mouth. For brain and other CNS tumours, chemotherapy can also be given directly into the cerebrospinal fluid through a ventricular access catheter, which is inserted during an operation. This device has a small tube that passes into the area of the brain where the cerebrospinal fluid flows. The treatment team can inject chemotherapy medicines directly into it, and also take samples of fluid out of it to run tests.

Radiation therapy

Radiation therapy (also called radiotherapy) uses high-energy X-rays or other types of radiation to destroy cancer cells or stop them from growing.

Radiation therapy may be used:





- after surgery (to destroy any remaining cancer cells)
- as the main treatment (if surgery is not a good option or chemotherapy does not stop the tumour growing)
- to relieve symptoms of the tumour.

Children and adults may have the same total dose of radiation to treat brain and other CNS tumours, but children may have smaller doses spread over a few days. Depending on where the tumour is and whether it has spread, high doses of radiation can be aimed precisely at the tumour, which limits the amount of radiation to other areas of the body. If a tumour has spread or has the potential to spread throughout the CNS, radiation may be given to the entire brain and spinal column.

Radiation therapy is not commonly used to treat brain and other CNS tumours in children under 3 years of age because it can have long-term side effects on developing brains. Young children usually have chemotherapy instead, and may have radiation therapy once they are old enough. If radiation therapy is included in your child's treatment, special care will be taken to reduce the risks.

Radiation therapy can affect your child's learning abilities. They will have regular assessments for learning disabilities throughout their childhood.

Targeted therapy

Some medicines can target the specific changes in cancer cells that make them different from normal cells. This means that they work differently from standard chemotherapy, and they usually have fewer side effects, or the side effects are not as severe.

A medicine called everolimus can be used for a particular type of brain and CNS tumour called subependymal giant cell astrocytoma (SEGA) to shrink it or slow its growth. It is taken as a tablet once a day.

Monoclonal antibodies (such as bevacizumab) are given intravenously, and can be used to kill cancer cells, slow their growth, or deliver medication directly to the cancer site.

Targeted therapies are usually given in combination with other treatments such as chemotherapy or surgery. Many targeted therapies are experimental and are only used in clinical trials.

Other treatments

Some treatments do not treat the tumour itself, but can help relieve symptoms that result from the tumour or the treatment. Common treatments of this kind include:

- corticosteroids, which can help reduce swelling in the skull, and reduce headaches, nausea and vomiting
- medicines that can reduce the chance of your child having seizures





- hormones that can restore your child's hormone balance if the tumour or the treatment has affected their pituitary gland (which controls hormone levels in the body).

Careful observation

If your child has a tumour that is not growing or spreading, they might be monitored closely but not given any treatment until they develop symptoms, or until their symptoms change. Careful observation is sometimes used for some low-grade gliomas.

Support

Diagnosis of cancer in a child is a very difficult time for the child, their family and their friends. You might feel overwhelmed, scared, anxious or angry. These are all normal feelings. It is very important to seek support from family, friends, health professionals or other services to help you, your child and your family cope with cancer.

Talk to your child's treatment team if you are having difficulties coping.

[Living with children's cancer](#) has information about physical, emotional and practical issues during and after diagnosis and treatment. There is also a page with helpful links on [where to find support](#).

The [Cancer Council](#) in your state or territory can give you general information about cancer, as well as information on resources and support groups in your local area. Call the Cancer Council Helpline from anywhere in Australia for the cost of a local call on **13 11 20**.

For additional specific information about childhood cancer, contact any of the major [children's hospitals and networks](#) in your state or territory.

Chance of cure

Many children with cancer are cured of the disease. Children's bodies have great capacity for healing. Also, huge improvements have been made in the treatment of childhood cancer in the past few decades. In the 1980s, around 65% of children diagnosed with cancer were alive more than 5 years





after their diagnosis. Today, around 83% of children are successfully treated and become long-term survivors.

Long-term survival (also called the outlook or prognosis) and treatment options depend on a range of factors, including:

- age of your child at diagnosis
- extent or stage of the cancer
- appearance of the cancer cells under the microscope (the shape, function and structure of the cells)
- how the cancer responds to treatment
- cancer or tumour biology, which includes
 - the patterns of the cancer cells
 - how different the cancer cells are from normal cells
 - how fast the cancer cells are growing.

Talk to your child's doctor about your child's individual disease, treatment options and outlook.

Clinical trials

Researchers are trialling new ways to diagnose and treat different types of cancer. Your child may be invited to be part of a clinical trial to test new ways of treating brain and other CNS tumours.

New treatments have to go through very strict regulation and approval processes before they can be used in a clinical trial. Your child's doctor will explain everything about the trial and give you detailed written information. You will need to give special permission for your child to be part of the trial.

Participating in a clinical trial may or may not directly benefit your child, but the results of clinical trials today will help children with cancer in the future.

See [Clinical trials and research](#) for more information, including whether there are any clinical trials your child can join.

More information

For more information about specific types of childhood brain and other CNS tumours, see:



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- [Brain and spinal cord tumors in children](#), from the American Cancer Society
- Treatment PDQs® from the National Cancer Institute (United States), including
 - [Childhood astrocytomas](#)
 - [Childhood brain and spinal cord tumors](#)
 - [Childhood brain stem glioma](#)
 - [Childhood CNS atypical teratoid/rhabdoid tumour](#)
 - [Childhood CNS embryonal tumours](#)
 - [Childhood CNS germ cell tumours](#)
 - [Childhood craniopharyngioma](#)
 - [Childhood ependymoma](#)

