

Central Nervous System (brain and spinal cord) Tumours

Information for people with cancer

This information should not be used to diagnose yourself or in place of a doctor's care.

The brain and spinal cord make up the central nervous system (CNS).

Brain

- The brain is a complex organ made up of nerve cells (neurons) and supporting tissues (glia).
- It has four main parts:
 - **Meninges** are the membranes that surround your brain.
 - **Cerebrum** is the largest part of your brain. It is split into two hemispheres (two halves) that each have multiple lobes (sections). Each lobe has specific functions.
 - **Cerebellum** is the back part of your brain, under your cerebrum.
 - **Brain stem** connects to your spinal cord
- The brain controls everything that goes on in your body: your senses, thought, reasoning, memory, emotion, movement, and breathing. It is also the root of your personality and behaviour.
- The adult brain weighs about 1.4 kg (3 lbs). The skull surrounds and protects the brain.

Spinal cord

- The spinal cord is a thin tube of nerve tissue that runs down your back from your brain into your pelvis.
- It is protected by bony vertebrae (spine).
- It carries nerve signals back and forth between your brain and the rest of your body.

Image of brain and spinal cord: visualsonline.cancer.gov/details.cfm?imageid=4279

Diagnosis and Staging

What are the signs and symptoms of CNS tumours?

Many of the common symptoms of central nervous system (CNS) tumours can also be caused by other conditions.

These are some symptoms of CNS tumours:

- Headaches that do not get better with usual headache treatments and are often worse in the morning.
- Seizures.
- Nausea (feeling queasy) and vomiting.
- Vision changes such as double vision (seeing two of everything) or loss of vision.
- Unusual or sudden changes in activity level, personality or behavior.
- Unusual drowsiness (very sleepy).
- Walking, balance or fine motor problems (for example, difficulty picking up a pen with your fingers).

If you have any symptoms that you are worried about, please talk to your family doctor or nurse practitioner.

How are CNS tumours diagnosed?

Tests that may help diagnose CNS tumours include:

- **Physical exam**
- **Neurological exam:** a doctor or nurse practitioner evaluates your nervous system, often using lights and reflex hammers, and asking questions.

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- **Imaging tests such as Magnetic Resonance Imaging (MRI) and Computerized tomography (CT):** For these scans, you are injected with a contrast agent which makes the tumour easier to see in the brain and spine.
- **Lumbar puncture (spinal tap):** For some tumours, a needle can be placed into your spinal canal to collect cerebrospinal fluid. This fluid can be tested for tumour cells.
- **Angiography:** a contrast agent is injected into your vein or artery. X-rays are then used to see the vein or artery. This test is used to look at the flow of blood in the brain.
- **Biopsy:** A small amount of tissue from the tumour is removed so that a specialist (pathologist) can examine it under the microscope and make a diagnosis.

For more information on tests used to diagnose cancer, see our Screening and Diagnosis pathfinder: bccancer.libguides.com/pathfinder-screening

What are the types of CNS tumours?

CNS tumours are sorted into types based on the type of brain cell involved and the grade of the tumour.

Grade of the tumour: How fast a tumour is growing determines the grade of the tumour. Brain tumours may have cells of different grades. The overall grade of the tumour is the highest grade of cell found in the tumour. Knowing the type helps us plan your treatment. It also tells us how your tumour might respond to treatment and whether it is more likely to come back after treatment.

CNS tumours have a grade from 1 to 4 based on how the cells look and behave.

- **Grade 1:** Slowest growing. Cells look almost normal. These tumours may be completely removed by surgery if they are in an area the surgeon can safely get to.

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- **Grade 2:** Grow slightly faster than grade 1. May infiltrate, meaning they can grow into tissues around them. Sometimes they come back (recur) after surgery. They can become more aggressive over time.
- **Grade 3:** Fast-growing and malignant (may also be called aggressive). Grow into surrounding tissue and often come back after surgery.
- **Grade 4:** Fast-growing and malignant. Cells look very different from normal brain cells and grow aggressively into surrounding tissues.

Staging

Staging tells us how much the cancer has spread throughout the body. CNS tumours do not usually spread to other parts of the body. Therefore, we do not stage CNS tumours.

Types of CNS tumours

Primary fast-growing, malignant tumours

Gliomas are tumours of the glia (supportive tissues).

- Astrocytes, oligodendrocytes and ependymal cells are different types of glial cells. The tumour types are astrocytomas, oligodendrogliomas and ependymomas. Oligodendrogliomas are particularly sensitive to treatment.
- Gliomas make up 45% (45 out of 100) of all tumours that start in brain cells (primary brain tumours).

Medulloblastoma is the most common malignant brain tumour in children, but can sometimes occur in adults. These start in the cerebellum, but they can spread to the spinal cord through the cerebrospinal fluid. We can test the tumour to see what type of medulloblastoma it is.

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Primary CNS lymphoma is a rare form of non-Hodgkin lymphoma that starts in the lymph tissue (part of the immune system) of the brain, spinal cord or meninges.

Primary low grade (slow-growing) tumours

- **Acoustic neuroma** (also called acoustic schwannoma): Tumour of the nerve leading from the inner ear to the brain. They are rare and most often seen in middle-aged women [See note, Statistics]. As the neuroma grows, it puts pressure on nearby brain structures.
- **Meningioma**: Tumour of the meninges (membranes). Most common CNS tumour. A person with this tumour often has no symptoms. Most are not aggressive but some are.
- **Pituitary adenoma**: Tumour of the pituitary gland. This gland is the master gland of the body. It produces hormones that control many processes in the body, like growth and reproduction.
- **Craniopharyngioma**: Tumour in the pituitary gland and nearby areas. These are not common.
- **Pineal gland tumour**: Rare. These may affect the production of melatonin (a hormone that controls the sleep-wake cycle).
- **Neurofibroma**: Tumour that grows in cells that support peripheral nerves (nerves that connect the brain and spinal cord to the body).

Secondary (metastatic) tumours

These tumours have spread to the brain from cancer that started somewhere else in the body. The original cancer is often in the lungs, breasts, colon, or skin (melanomas). Sometimes we cannot tell where the original cancer started. Secondary brain tumours are more common than primary brain tumours, which start in the brain.

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Leptomeningeal tumours grow in the membranes covering the brain and spinal cord. They often spread from a breast cancer.

Molecular characteristics of CNS tumours

Certain features of the genetic makeup of the cells in the tumour are as important as the way the cells look under a microscope. These genetic, or molecular, features of cells help us understand how a tumour will behave. They also help us plan the right treatments for people with brain tumours. Some of the important molecular features of tumours are:

MGMT (methylguanine methyltransferase): MGMT is a gene that repairs damaged DNA in our cells. Under normal circumstances this is a good thing. However, cancer treatments work by damaging tumour cell DNA. So MGMT can help a cancer cell survive treatments like radiation therapy or chemotherapy. If MGMT is methylated (or silenced), it does not work properly. It cannot undo the damage done to cancer cells by treatment. Glioblastoma tumours that are MGMT methylated are more likely to respond well to treatment. If an older person has a tumour that is not MGMT methylated, we may not give them aggressive chemotherapy, because in these cases, the treatment can be more harmful than helpful.

IDH (isocitrate dehydrogenase) 1 and 2: People with tumours that have mutations (changes) in the IDH gene may respond better to treatment and have better outcomes.

1p/19q (short arm of chromosome 1 and long arm of chromosome 19): Cancer cells that form oligodendrogliomas often have missing genetic material from chromosomes 1 and 19. These tumours are often more sensitive to treatment and associated with long survival for patients.

Treatment

How are CNS tumours treated?

Treatment may be different for each person, depending on their health and their tumour type. Your treatment may include:

Surgery

- The goal of surgery is to remove as much of the tumour as possible without damaging critical parts of your brain or spinal cord. Sometimes only a very small piece of the tumour can be safely removed. This piece of tissue is called a biopsy. It is tested to diagnose the tumour and plan treatment.
- For brain surgery, a craniotomy is almost always done. A piece of your skull bone is removed and the bone is put back at the end of the surgery.
- If a grade 1 tumour can be completely removed, this may be the only treatment needed.
- Sometimes surgical “debulking” is done to help relieve pressure from fluid buildup or a large tumour. When some of the tumour is taken out, it can help relieve symptoms.

Radiation therapy (high energy x-rays that kill or shrink cancer cells)

- Sometimes given after surgery to slow or stop the growth of remaining tumour cells.
- Radiation therapy is given using very specialized techniques to focus the beam of radiation at the tumour and minimize radiation of nearby tissues. For gliomas (see types of CNS tumours, above), we also give radiation to a small area around the tumour, to treat cells that may have spread there.
- Short-term side effects can include fatigue (extreme tiredness), hair loss, skin reactions, headache, nausea (feeling queasy).

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- Longer term side effects can include problems with memory and thinking, and lower hormone production.
- For more information about radiation therapy go to:
bccancer.bc.ca/our-services/treatments/radiation-therapy

Systemic therapy (drug therapy or chemotherapy)

- Chemotherapy pills and intravenous (through your vein) chemotherapies are sometimes given after surgery and radiotherapy. We do this to kill any remaining cancer, to lower the risk of the cancer coming back (recurrence), or to treat a tumour that does start to grow again (recurs) after it has previously been treated,
- Sometimes we give chemotherapy with radiation therapy to make the radiation work better.
- Other drugs may be used to help manage symptoms. For example, steroids, such as dexamethasone, can help with swelling in the brain. Also, seizure medications can prevent seizures, help make them less severe and reduce how often they happen.
- For more information about systemic therapy go to:
[bccancer.bc.ca/our-services/treatments/systemic-therapy-\(chemotherapy\)](http://bccancer.bc.ca/our-services/treatments/systemic-therapy-(chemotherapy))

What is the follow-up after treatment?

- Follow-up testing and appointments are based on your disease type.
- Please read our Follow-up Plan for Brain Tumour Patients: www.bccancer.bc.ca/cancer-management-guidelines-site/Documents/2015_Follow-up%20prog%20forBrainTumourPatients.pdf

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- Guidelines for follow-up after treatment for CNS tumours are on our website: www.bccancer.bc.ca/health-professionals/clinical-resources/cancer-management-manual/neuro-oncology/neuro-oncology
- These recommendations written for your doctor, nurse practitioner or specialist. You can look at them to see what appointments and tests you might need after treatment.
- After treatment, you may return to the care of your family doctor or specialist for regular follow-up. If you do not have a family doctor, please talk to your BC Cancer health care team.
- The BC Cancer Life after Cancer page has information on issues that cancer survivors may face: bccancer.bc.ca/lifeaftercancer

More Information

What causes CNS tumours and who gets them?

CNS tumours are rare. They make up about 2% (2 out of 100) of cancers. They are most common in young children and older adults, but can occur at any age.

Unlike other cancers, there are few known risk factors for CNS tumours.

Exposure to radiation, either from environmental sources or from radiation treatment, can increase your risk of CNS tumours. However, the benefit of treating an existing life-threatening tumour with radiation outweighs the potential risk of developing a tumour later in life.

There are some genetic conditions that may increase your risk of a brain tumour. These are not very common. They include neurofibromatosis types 1 and 2, Li Fraumeni syndrome, Lynch syndrome and tuberous sclerosis.

Statistics on CNS tumours

- B.C. statistics: www.bccancer.bc.ca/health-info/disease-system-statistics/statistics-by-cancer-type

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- Canada statistics: www.cancer.ca/en/cancer-information/cancer-type/brain-spinal/statistics/?region=bc

Note: Available statistics do not have information about the inclusion of transgender and gender diverse participants. It is unknown how these statistics apply to transgender and gender diverse people. Patients are advised to speak with their primary care provider or specialists about their individual considerations and recommendations.

Can I help prevent CNS tumours?

We do not know what causes most primary brain tumours so we do not know how to prevent them. The only way we know of is to avoid unnecessary radiation exposure.

Secondary brain tumours from lung cancer are common and lung cancer is strongly related to tobacco use.

- To lower your risk, do not smoke and try not to be exposed to tobacco and cigarette smoke. Even if you have been using tobacco for many years, quitting will lower your cancer risk. Support is available to help you successfully quit. Visit the BC Cancer Tobacco and Cancer Prevention page: www.bccancer.bc.ca/health-info/prevention/tobacco

Is there screening for CNS tumours?

There is no screening program for these tumours.

Where can I find more information?

- If you have questions about CNS tumours, please talk to your health care team.
- Our librarians can help you find the information you need. Visit our Library page: bccancer.bc.ca/our-services/services/library

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- BC Cancer Library Brain and Central Nervous System Cancer pathfinder: bccancer.libguides.com/pathfinder_brain
- Managing Your Symptoms: www.bccancer.bc.ca/health-info/coping-with-cancer/managing-symptoms-side-effects
- Emotional Support: www.bccancer.bc.ca/health-info/coping-with-cancer/emotional-support