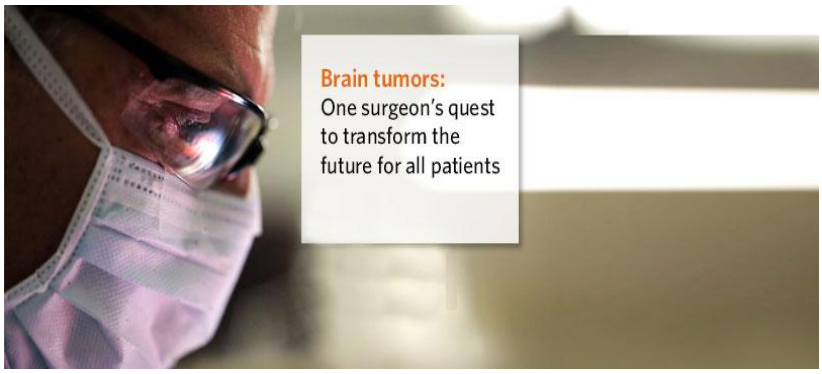
# Brain and Spinal Cord Tumours in Adults


## WHAT IS BRAIN/CNS TUMORS IN ADULTS?

The body is made up of trillions of living cells. Normal body cells grow, divide to make new cells, and die in an orderly way. During the early years of a person's life, normal cells divide faster to allow the person to grow. After the person becomes an adult, most cells divide only to replace worn-out or dying cells or to repair injuries.

Cancer begins when cells in a part of the body start to grow out of control. There are many kinds of cancer, but they all start because of out-of-control growth of abnormal cells.

Cancer cell growth is different from normal cell growth. Instead of dying, cancer cells continue to grow and form new, abnormal cells. In most cases the cancer cells form a tumor. Cancer cells can also invade (grow into) other tissues, something that normal cells cannot do. Growing out of control and invading other tissues are what makes a cell a cancer cell.

Cells become cancer cells because of damage to DNA. DNA is in every cell and directs all its actions. In a normal cell, when DNA is damaged the cell either repairs the damage or the cell dies. In cancer cells, the damaged DNA is not repaired, but the cell doesn't die like it should. Instead, this cell goes on making new cells that the body doesn't need. These new cells will all have the same damaged DNA as the first abnormal cell does.

People can inherit damaged DNA, but most often the DNA damage is caused by mistakes that happen while the normal cell is reproducing or by something in our environment. Sometimes the cause of the DNA damage is something obvious, like cigarette smoking. But often no clear cause is found.

Cancer cells often travel to other parts of the body, where they begin to grow and form new tumors that replace normal tissue. This process is called *metastasis*. It happens when the cancer cells get into the bloodstream or lymph vessels of our body.

No matter where a cancer may spread, it is named (and treated) based on the place where it started. For example, breast cancer that has spread to the liver is still breast cancer, not liver cancer. Likewise, prostate cancer that has spread to the bone is still prostate cancer, not bone cancer.

Different types of cancer can behave very differently. They grow at different rates and respond to different treatments. That is why people with cancer need treatment that is aimed at their particular kind of cancer.

Not all tumors are cancerous. Tumors that aren't cancer are called *benign*. Benign tumors cannot grow into (invade) other tissues or spread to other parts of the body (metastasize). In most parts of the body, these tumors are almost never life threatening. But in the brain, even benign tumors can sometimes grow large enough to damage nearby normal brain tissue. This can be disabling and might even be life threatening in some cases.

## CAUSES, RISK FACTORS, AND PREVENTION

### What are the risk factors for brain and spinal cord tumors?

A risk factor is anything that affects your chance of getting a disease such as a brain or spinal cord tumor. Different types of cancer have different risk factors. Some risk factors, like smoking, you can change. Others, like your age or family history, can't be changed.

But having a risk factor, or even several, does not always mean that a person will get the disease, and many people get tumors without having any known risk factors. Even if a person has a risk factor, it is often very hard to know how much it contributed to the tumor.

Most brain tumors are not linked with any known risk factors and have no obvious cause. But there are a few factors that can raise the risk of brain tumors.

### Radiation exposure

The best known environmental risk factor for brain tumors is radiation exposure, most often from radiation therapy to treat some other condition. For example, before the risks of radiation were known, children with ringworm of the scalp (a fungal infection) were sometimes treated with low-dose radiation therapy, which was later found to increase their risk of brain tumors as they got older.

Today, most radiation-induced brain tumors are caused by radiation to the head given to treat other cancers. They occur most often in people who received radiation to the brain as children as part of their treatment for leukemia. These brain tumors usually develop around 10 to 15 years after the radiation.

Radiation-induced tumors are still fairly rare, but because of the increased risk (as well as the other side effects), radiation therapy to the head is only given after carefully weighing the possible benefits and risks. For most patients with other cancers involving the brain or head, the benefits of radiation therapy far outweigh the risk of developing a brain tumor years later.

The possible risk from exposure to imaging tests that use radiation, such as x-rays or CT scans, is not known for sure. These tests use much lower levels of radiation than those used in radiation treatments, so if there is any increase in risk, it is likely to be very small. But to be safe, most doctors recommend that people (especially children and pregnant women) not get these tests unless they are clearly needed.

**Family history**

Most people with brain tumors do not have a family history of the disease, but in rare cases brain and spinal cord cancers run in families. In general, patients with familial cancer syndromes tend to have many tumors that first occur when they are young. Some of these families have well-defined disorders, such as:

**Neurofibromatosis type 1 (NF1)**

This genetic disorder, also known as *von Recklinghausen disease*, is the most common syndrome linked to brain or spinal cord tumors. People with this condition have higher risks of schwannomas, meningiomas, and certain types of gliomas, as well as neurofibromas (benign tumors of peripheral nerves). Changes in the *NF1* gene cause this disorder. These changes are inherited from a parent in about half of all cases. In the other half, the *NF1* gene changes occur before birth in people whose parents did not have this condition.

**Neurofibromatosis type 2 (NF2)**

This condition, which is much less common than NF1, is associated with vestibular schwannomas (acoustic neuromas), which almost always occur on both sides of the head. It is also linked with an increased risk of meningiomas or spinal cord ependymomas. Changes in the *NF2* gene are responsible for neurofibromatosis type 2. Like *NF1*, the gene changes are inherited in about half of cases or may occur before birth in children without a family history.

**Tuberous sclerosis**

People with this condition may have subependymal giant cell astrocytomas (SEGAs), which are low-grade astrocytomas that develop beneath the ependymal cells of the ventricles). They may also have other benign tumors of the brain, skin, heart, kidneys, and other organs. This condition is caused by changes in either the *TSC1* or the *TSC2*gene. These gene changes can be inherited from a parent, but most often they develop in people without a family history.

**Von Hippel-Lindau disease**

People with this condition tend to develop benign or cancerous tumors in different parts of the body, including hemangioblastomas (blood vessel tumors) in the brain, spinal cord, or retina, as well as tumors of the inner ear, kidney, adrenal gland, and pancreas. It is caused by changes in the *VHL* gene. Most often the gene changes are inherited, but in some cases the changes happen before birth in people whose parents don't have them.

**Li-Fraumeni syndrome**

People with this condition are at higher risk for developing gliomas, along with breast cancer, soft tissue sarcomas,leukemia, and adrenal gland cancer, and certain other types of cancer. It is caused by changes in the *TP53* gene.

**Other syndromes**

Other inherited conditions are also linked with increased risks of certain types of brain and spinal cord tumors, including:

- Gorlin syndrome (basal cell nevus syndrome)
- Turcot syndrome
- Cowden syndrome

Some families may have genetic disorders that are not well recognized or that may even be unique to a particular family.

**Immune system disorders**

People with impaired immune systems have an increased risk of developing lymphomas of the brain or spinal cord (known as *primary CNS lymphomas*). Lymphomas are cancers of lymphocytes, a type of white blood cell that fights disease. Primary CNS lymphoma is less common than lymphoma that develops outside the brain.

A weakened immune system can be congenital (present at birth), or it can be caused by treatments for other cancers, treatment to prevent rejection of transplanted organs, or diseases such as the acquired immunodeficiency syndrome(AIDS).

**Factors with uncertain, controversial, or unproven effects on brain tumor risk**

**Cell phone use**

This has been the subject of a great deal of debate in recent years. Cell phones give off radiofrequency (RF) rays, a form of energy on the electromagnetic spectrum between FM radio waves and those used in microwave ovens, radar, and satellite stations. Cell phones do not give off ionizing radiation, the type that can cause cancer by damaging the DNA inside cells. Still, there have been concerns that the phones, whose antennae are built-in and therefore are placed close to the head when being used, might somehow raise the risk of brain tumors.

Some studies have suggested a possible increased risk of brain tumors or of vestibular schwannomas with cell phone use, but most of the larger studies done so far have not found an increased risk, either overall or among specific types of tumors. Still, there are very few studies of long-term use (10 years or more), and cell phones haven't been around long enough to determine the possible risks of lifetime use. The same is true of any possible higher risks in children, who are increasingly using cell phones. Cell phone technology also continues to change, and it's not clear how this might affect any risk.

These risks are being studied, but it will probably be many years before firm conclusions can be made. In the meantime, for people concerned about the possible risks, there are ways to lower your exposure, such as using an earpiece to move the phone itself away from the head. For more information, see our document *Cellular Phones*.

**Other factors**

Other environmental factors such as exposure to vinyl chloride (a chemical used to manufacture plastics), petroleum products, and certain other chemicals have been linked with an increased risk of brain tumors in some studies but not in others.

Exposure to aspartame (a sugar substitute), exposure to electromagnetic fields from power lines and transformers, and infection with certain viruses have been suggested as possible risk factors, but most researchers agree that there is no convincing evidence to link these factors to brain tumors. Research on these and other potential risk factors continues.

## EARLY DETECTION, DIAGNOSIS, AND STAGING

**Can brain and spinal cord tumors in adults be found early?**

At this time there are no widely recommended tests to screen for brain and spinal cord tumors. (Screening is testing for cancer in people without any symptoms.) Most brain tumors are found when a person sees a doctor because of signs or symptoms they are having (see "Signs and symptoms of adult brain and spinal cord tumors").

Most often, the outlook for people with brain and spinal cord tumors depends on their age, the type of tumor, and its location, not by how early it is detected. But as with any disease, earlier detection and treatment is likely to be helpful.

For people with certain inherited syndromes that put them at higher risk for brain tumors, such as neurofibromatosis or tuberous sclerosis, doctors often recommend frequent physical exams and other tests starting when they are young. In some cases these tests can find tumors when they are still small. Not all tumors related to these syndromes may need to be treated right away, but finding them early might help doctors monitor them so that they can be treated quickly if they begin to grow or cause problems.

## TREATING BRAIN/CNS TUMORS IN ADULTS

**How are brain and spinal cord tumors in adults treated?**

*This information represents the views of the doctors and nurses serving on the American Cancer Society's Cancer Information Database Editorial Board. These views are based on their interpretation of studies published in medical journals, as well as their own professional experience.*

*The treatment information in this document is not official policy of the Society and is not intended as medical advice to replace the expertise and judgment of your cancer care team. It is intended to help you and your family make informed decisions, together with your doctor.*

*Your doctor may have reasons for suggesting a treatment plan different from these general treatment options. Don't hesitate to ask him or her questions about your treatment options.*

**General comments about treatment**

Brain and spinal cord tumors can often be hard to treat and may require care from a team of different types of doctors. This team is often led by a neurosurgeon, a doctor who uses surgery to treat brain and nervous system tumors. Other doctors on the team may include:

- Neurologist: a doctor who diagnoses brain and nervous system diseases and treats them with medicines
- Radiation oncologist: a doctor who uses radiation to treat cancer
- Medical oncologist: a doctor who uses chemotherapy and other medicines to treat cancers
- Endocrinologist: a doctor who treats diseases in glands that secrete hormones

Many other specialists may be involved in your care as well, including physician assistants, nurse practitioners, nurses, psychologists, social workers, rehabilitation specialists, and other health professionals.

Several types of treatment can be used to treat brain and spinal cord tumors, including:

- Surgery
- Radiation therapy
- Chemotherapy
- Targeted therapy
- Other types of drugs

Treatment is based on the type of tumor and other factors, and often more than one type of treatment is used. Doctors plan each person's treatment individually to give them the best chance of treating the cancer while limiting the side effects as much as possible.

It's important to discuss all of your treatment options as well as their possible side effects with your treatment team to help make the decision that best fits your needs. If there is anything you don't understand, ask to have it explained. (See the section "What should you ask your doctor about adult brain and spinal cord tumors?" for some questions to ask.)

If time permits, getting a second opinion from a doctor experienced with your type of tumor is often a good idea. It can give you more information and help you feel more confident about the treatment plan you choose.

The next few sections describe the various types of treatments used for brain and spinal cord tumors. This is followed by a description of the most common approaches used based on the type of tumor.

## TALKING WITH YOUR DOCTOR

### What should you ask your doctor about adult brain and spinal cord tumors?

It's important for you to have honest, open discussions with your cancer care team. Feel free to ask any question, no matter how small it might seem. Here are some questions you might want to ask. Be sure to add your own questions as you think of them. Nurses, social workers, and other members of the treatment team can also answer many of your questions.

- What kind of tumor do I have?
- Is the tumor benign or malignant? What does this mean?
- Where in the brain or spinal cord is the tumor and how far has it spread?
- Do I need other tests before we can decide on treatment?
- How much experience do you have treating this type of tumor?
- Should I get a second opinion? Can you recommend a doctor or cancer center?
- What are my treatment choices? What do you recommend? Why?
- What's the goal of treatment (cure, prolonging life, relieving symptoms, etc.)?
- Will treatment relieve any of the symptoms I now have?
- What are the possible risks or side effects of treatment? What disabilities might I develop?
- What should I do to be ready for treatment?
- How long will treatment take? What will it be like? Where will it be given?
- What is my expected prognosis (outlook)?
- What would we do if the treatment doesn't work or if the tumor comes back?
- What type of follow-up will I need after treatment?
- Where can I find more information and support?

Along with these sample questions, be sure to write down any others you want to ask. For instance, you might want information about recovery times so that you can plan your work or activity schedule. Or you might want to ask aboutclinical trials that might be right for you.

## AFTER TREATMENT

### What happens after treatment for adult brain and spinal cord tumors?

For some people with brain or spinal cord tumors, treatment can remove or destroy the tumor. Completing treatment can be both stressful and exciting. You may be relieved to finish treatment, but find it hard not to worry about the tumor coming back. (When a tumor comes back after treatment, it is called *recurrence*.) This is a very common concern in people who have had a brain or spinal cord tumor.

It may take a while before your fears lessen. But it may help to know that many cancer survivors have learned to live with this uncertainty and are leading full lives. Our document *Living With Uncertainty: The Fear of Cancer Recurrence*, gives more detailed information on this.

For other people, the tumor may never go away completely. Some people may continue to be treated with radiation therapy, chemotherapy, or other treatments to try to keep the tumor in check. Learning to live with a tumor that does not go away can be difficult and very stressful. It has its own type of uncertainty. Our document *When Cancer Doesn't Go Away* talks more about this.

**Follow-up care**

If you have completed treatment, your doctors will still want to watch you closely. It's very important to keep all follow-up appointments. During these visits, your doctors will ask about symptoms, examine you, and may order lab tests or imaging tests such as MRI scans to look for a recurrence of the tumor. In some cases, some of the tumor may still be left behind after treatment. Even tumors that have been treated successfully can sometimes come back.

Whether the tumor was removed completely or not, your health care team will want to follow up closely with you, especially in the first few months and years after treatment to make sure there is no progression or recurrence. Depending on the type and location of the tumor and the extent of the treatment, the team will decide which tests should be done and how often.

During this time, it is important to report any new symptoms to your doctor right away, so the cause can be found and treated, if needed. Your doctor can give you an idea of what to look for. If you need further treatment at some point, the doctor will go over your options with you.

Should your tumor come back, our document *When Your Cancer Comes Back: Cancer Recurrence* can give you information on how to manage and cope with this phase of your treatment.

## WHAT`S NEW IN BRAIN/CNS TUMORS IN ADULTS RESEARCH?

**What's new in adult brain and spinal cord tumor research and treatment?**

There is always research going on in the area of brain and spinal cord tumors. Scientists are looking for causes and ways to prevent these tumors, and doctors are working to improve treatments.

**Genetics**

Researchers are looking for changes inside brain tumor cells to see if they can be used to help guide treatment. For example, doctors have found that patients with oligodendrogliomas whose cells are missing parts of certain chromosomes (known as a *1p19q co-deletion*) are much more likely to be helped by chemotherapy than patients whose tumors do not.

**Imaging and surgery techniques**

Recent advances have made surgery for brain tumors much safer and more successful. Some of these newer techniques include:

- Functional magnetic resonance imaging (fMRI, described in "How are brain and spinal cord tumors in adults diagnosed?"). This technique can help identify important functional areas of the brain and how close they are to the tumor.
- Magnetic resonance spectroscopic imaging (MRSI, described in "How are brain and spinal cord tumors in adults diagnosed?). In this approach, specially processed MRS information is used to make a map of important chemicals involved in tumor metabolism. This is being developed to help surgeons direct their biopsies to the most abnormal areas in the tumor and to help doctors direct radiation and evaluate the effects of chemotherapy or targeted therapy.
- Fluorescence-guided surgery. For this approach, the patient drinks a special fluorescent dye a few hours before surgery. The dye is taken up mainly by the tumor, which then glows

when the surgeon looks at it under special lighting from the operating microscope. This lets the surgeon better separate tumor from normal brain tissue.

- Newer surgical approaches for some types of tumors. For example, a newer approach to treat some tumors near the pituitary gland is to use an endoscope, a thin tube with a tiny video camera lens at the tip. The surgeon passes the endoscope through a small hole made in the back of the nose to operate through the nasal passages, limiting the potential damage to the brain. A similar technique can be used for some tumors in the ventricles, where a small opening in the skull near the hairline serves as the point of endoscope insertion. The use of this technique is limited by the tumor's size, shape, and position.

**Radiation therapy**

Some newer types of external radiation therapy let doctors deliver radiation more precisely to the tumor, which helps spare normal brain tissue. Techniques such as 3-dimensional conformal radiation therapy (3D-CRT), intensity modulated radiation therapy (IMRT), and proton beam therapy are described in the section "Radiation therapy for adult brain and spinal cord tumors."

Newer methods of treatment planning are also being studied. For example, image-guided radiation therapy (IGRT) uses a CT scan done just before each treatment to better guide the radiation to its target.

**Chemotherapy**

Along with developing and testing new chemotherapy drugs, many researchers are testing new ways to get chemotherapy to the brain tumor.

Many chemotherapy drugs are limited in their effectiveness because the tightly controlled openings in the brain capillaries, sometimes referred to as the *blood-brain barrier*, prevents them from getting from the bloodstream to the brain. Researchers are now trying to modify some of these drugs by putting them in tiny droplets of fat (liposomes) or attaching them to molecules that normally cross the blood-brain barrier, to help them work better. This is an area of active research and clinical trials.

For another newer method called *convection-enhanced delivery*, a small tube is placed into the tumor in the brain through a small hole in the skull during surgery. The tube extends through the scalp and is connected to an infusion pump, through which drugs can be given. This can be done for hours or days and may be repeated more than once, depending on the drug used. This method is still being studied in clinical trials.

**Other new treatment strategies**

Researchers are also testing some newer approaches to treatment that may help doctors target tumors more precisely. This could lead to treatments that work better and cause fewer side effects. Several of these treatments are still being studied.

**Tumor vaccines**

Several vaccines are being tested against brain tumor cells. Unlike vaccines against infections, these vaccines are meant to help treat the disease instead of prevent it. The goal of the vaccines is to stimulate the body's immune system to attack the brain tumor.

Early study results of vaccines to help treat glioblastoma have shown promise, but more research is needed to determine how well they work. At this time, brain tumor vaccines are available only through clinical trials.

**Angiogenesis inhibitors**

Tumors need to create new blood vessels (a process called *angiogenesis*) to keep their cells nourished. New drugs that attack these blood vessels are used to help treat some cancers. One of these drugs, bevacizumab (Avastin), has been approved by the FDA to treat recurrent glioblastomas because it has been shown to slow the growth of some tumors.

Other drugs that impair blood vessel growth, such as sorafenib (Nexavar) and trebananib, are being studied and are available through clinical trials.

**Growth factor inhibitors**

Tumor cells are often very sensitive to proteins called *growth factors*, which help them grow and divide. Newer drugs target some of these growth factors, which may slow the growth of tumor cells or even cause them to die. Several of these targeted drugs are already used for other types of cancer, and some are being studied to see if they will work for brain tumors as well.

**Hypoxic cell sensitizers**

Some drugs increase the oxygen content in tumors, which might make tumor cells more likely to be killed by radiation therapy if they are given before treatment. These types of drugs are now being studied to see if they can improve treatment outcomes.

**Electric treatment fields**

The NovoTTF-100A system is approved by the FDA to treat glioblastomas that are no longer responding to other treatments. To use this device, the head is shaved and 4 sets of electrodes are placed on the scalp. The electrodes are attached to a battery pack and are worn for most of the day. They generate mild electric currents that are thought to affect tumor cells in the brain more than normal cells. In a clinical trial, people using the device lived about as long as those getting further chemotherapy, although they reported a better quality of life because of fewer side effects.

**Source** American Cancer Society®