

About Brain Tumors

a primer for patients and caregivers



American
Brain Tumor
Association®

Providing and pursuing answers®

About the American Brain Tumor Association

Founded in 1973, the American Brain Tumor Association (ABTA) was the first national nonprofit organization dedicated solely to brain tumor research. The ABTA has since expanded our mission and now provides comprehensive resources to support the complex needs of brain tumor patients and caregivers, across all ages and tumor types, as well as the critical funding of research in the pursuit of breakthroughs in brain tumor diagnoses, treatments and care.

To learn more, visit abta.org.

We would like to thank the many healthcare professionals who have helped to keep this publication up-to-date throughout the years. The following are those who have graciously contributed their time in reviewing this version of the primer:

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This publication is not intended as a substitute for professional medical advice and does not provide advice on treatments or conditions for individual patients. All health and treatment decisions must be made in consultation with your physician(s), utilizing your specific medical information. Inclusion in this publication is not a recommendation of any product, treatment, physician or hospital.

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INTRODUCTION

Learning you or your loved one has a brain tumor can be very frightening.

You may know little about tumors and even less about the brain. You might be confused about the new terms you are hearing, angry because you need to make decisions you are not prepared for and dazed by all of the changes in your life.

As you begin this new path in life, please know that you are not alone. The American Brain Tumor Association (ABTA) is here to help you throughout this journey. This book was written to help you, your family and your friends learn more about brain tumors. We offer information and resources and share suggestions and experiences from patients and families who have lived with a brain tumor. We hope this knowledge will offer a degree of comfort and help you feel more in control of your life during this difficult time.

Our team of professionals can provide additional information about tumors, treatment and support resources. We also encourage you to visit our website at www.abta.org. For more information, please call our CareLine at 800-886-ABTA (2282) or send us an email at abtacares@abta.org.

Chapter 1: Brain Tumor Basics

The human body is made up of trillions of cells. Normally, the adult body only forms new cells when they are needed to replace old or damaged ones. Infants and children create new cells to complete their development in addition to replacing old or damaged ones. When cells begin to multiply when they are not needed, a tumor develops.

A brain tumor is a mass of unnecessary cells growing in the brain or central spine canal. There are two basic types of brain tumors – primary brain tumors and metastatic brain tumors. Primary brain tumors start and tend to stay in the brain. Metastatic brain tumors begin as cancer elsewhere in the body and spread to the brain.

> Each year more than 68,470 people in the United States are diagnosed with a primary brain tumor and more than twice that number are diagnosed with a metastatic tumor.

PRIMARY BRAIN TUMORS

Primary brain tumors start in the brain or central spinal canal and tend to not spread to other parts of the body. Brain tumors tend to be classified as either malignant or non-malignant.

Malignant brain tumors

Malignant brain tumors are made up of cells that appear abnormal when viewed under a microscope. These tumors tend to be rapid-growing, invasive and life-threatening. Malignant brain tumors are made up of cancerous cells.

Malignant brain tumors can spread within the brain and spine, but they rarely spread to other parts of the body. They lack distinct borders due to their tendency to send “roots” into nearby normal, healthy tissue. They can also shed cells that travel to distant parts of the brain and spine by way of the cerebrospinal fluid. Some malignant tumors, however, do remain localized to a region of the brain or spinal cord.

Non-malignant Tumors:

- *Slow growing*
- *Distinct borders*
- *Rarely Spread*

Malignant Tumors::

- *Usually rapid growing*
- *Invasive*
- *Life threatening*

Benign (non-malignant) brain tumors

Growths or tumors that are not cancerous are often referred to as benign. Brain tumors composed of benign cells can still be located in vital areas of the brain, making them life-threatening. For this reason, we refer to benign brain tumors as non-malignant because even though they are not “cancer,” they can still be a serious medical condition.

Non-malignant tumors consist of very slow growing cells that have an almost normal appearance when viewed under a microscope. They also usually have distinct borders and rarely spread. Some non-malignant tumors may recur (or progress), spread to another site, or cause significant symptoms (either from the tumor itself or from treatment).

Cancer is a disease defined by:

- *Unregulated growth of abnormal cells*
- *Abnormal cells that grow into or around parts of the body and interfere with their normal functioning*
- *Spread to distant organs in the body*

Brain tumors can be called malignant if they:

- *Have the characteristics of cancer cells*
- *Are located in a critical part of the brain*
- *Are causing life threatening damage*

METASTATIC BRAIN TUMORS

Cancer cells that begin growing elsewhere in the body and then travel to the brain form metastatic brain tumors. For example, cancers of the lung, breast, colon and skin (melanoma) frequently spread to the brain via the bloodstream or a magnetic-like attraction to other organs of the body.

All metastatic brain tumors are, by definition, malignant and can truly be called “brain cancer.”

TUMOR NAMES

Tumors are diagnosed, and then named, based on a classification system. Most medical centers now use the World Health Organization (WHO) classification system for this purpose.

TUMOR GRADING

A tumor grade is a way to classify a tumor's level of malignancy or aggressiveness and will help members of the healthcare team communicate more clearly about the tumor, determine treatment options, and predict outcomes.

Tumors are assigned Grade I, II, III, or IV based on abnormalities of the cells they contain. A tumor can have more than one grade of cell. The highest, or most malignant, grade of cell determines the tumor's grade, even if most of the tumor is made up of lower-grade cells.

Using the WHO grading system, grade I tumors are the least malignant and are usually associated with long-term survival. These tumors grow slowly and have an almost normal appearance when viewed through a microscope. Surgery alone might be an effective treatment for this grade of tumor.

Grade II tumors are relatively slow-growing and have a slightly abnormal microscopic appearance. Some can spread into nearby normal tissue and recur. Sometimes these tumors recur as a higher grade tumor.

Grade III tumors are malignant, although there is not always a sharp distinction between a grade II and a grade III tumor. The cells of a grade III tumor are actively reproducing abnormal cells, which grow into nearby normal brain tissue. These tumors tend to recur, often as a higher grade tumor.

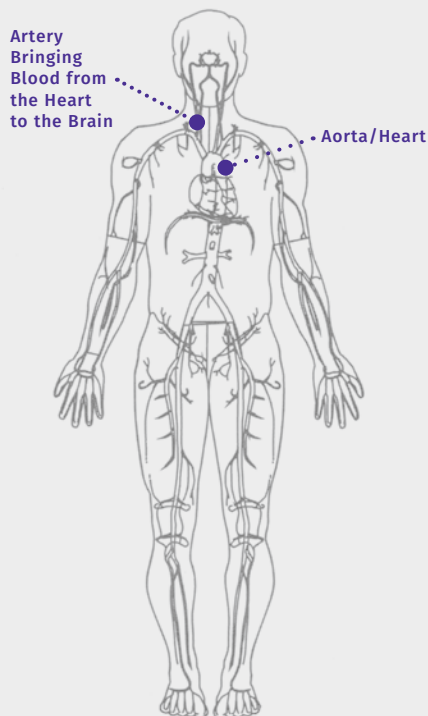
Grade IV tumors are the most malignant. They reproduce rapidly, can have a bizarre appearance when viewed under the microscope and easily grow into surrounding normal brain tissue. These tumors form new blood vessels so they can maintain their rapid growth. They also have areas of dead cells in their center. Glioblastoma is the most common example of a grade IV tumor.

Some tumors undergo change and a non-malignant growth might become malignant or, as previously mentioned, a lower-grade tumor might recur as a higher-grade tumor. Your doctor can tell you if your tumor might have this potential.

TUMOR ADVANCED ANALYSIS

As more brain tumors are being studied and researched, we now know that some tumors have additional characteristics that are more informative than just knowing the basic tumor type or the WHO grade. These characteristics may be the result of alterations of a cell receptor, a specific protein product, a mutation, or another molecular/genetic alteration.

ARTERIAL BLOOD FLOW



Grade is assigned based on the tumor's microscopic appearance using some or all of the following criteria:

- Similarity to normal cells (atypia)
- Rate of growth (mitotic index)
- Indications of uncontrolled growth
- Dead tumor cells in the center of the tumor (necrosis)
- Potential for invasion and/or spread (infiltration) based on whether or not it has a definitive margin (diffuse or focal)
- Blood supply (vascularity)

These characteristics are refining our historical understanding of tumor’s presentation, natural history, response to specific treatments, availability of certain treatments, availability of clinical trials, symptoms, and outcomes. The application of these characteristics to the “real-time” treatment decisions for individual patients is often called personalized medicine or precision medicine. Ask your doctor to discuss how advanced analysis may be able to help you or your loved one make the most informed decisions.

Grade I Tumor

- *Slow-growing cells*
- *Almost normal appearance under a microscope*
- *Least malignant*
- *Usually associated with long-term survival*

Grade II Tumor

- *Relatively slow-growing cells*
- *Slightly abnormal appearance under a microscope*
- *Can invade adjacent normal tissue*
- *Can recur as a higher grade tumor*

Grade III Tumor

- *Actively reproducing abnormal cells*
- *Abnormal appearance under a microscope*
- *Infiltrate adjacent normal brain tissue*
- *Tumor tends to recur, often as a higher grade*

Grade IV Tumor

- *Abnormal cells which reproduce rapidly*
- *Very abnormal appearance under a microscope*
- *Form new blood vessels to maintain rapid growth*
- *Areas of dead cells (necrosis) in center*

> Please ask a member of your healthcare team to complete the form on the next page. Use it to learn the exact spelling of your tumor type and its location, your medications, and resources for additional information.

THE NAME OF MY TUMOR IS:

- Astrocytoma grade I
- Astrocytoma grade II
- Astrocytoma grade III, also called Anaplastic Astrocytoma or Malignant Astrocytoma
- Ependymoma grade I, II, III
- Glioblastoma, also called Glioblastoma Multiforme or Astrocytoma grade IV
- Medulloblastoma
- Meningioma
- Metastatic tumor (primary site: _____)
- Oligodendroglioma
- Anaplastic Oligodendroglioma (III)
- Pituitary Adenoma, also called Pituitary Tumor
- Other: _____

ADVANCED ANALYSES OF MY TUMOR IS:

(e.g., BRAFv600E mutation status; MGMT methylation status; IDH mutation status)

MY MEDICAL TEAM CONTACT IS:

Program/Office name: _____

Phone: _____

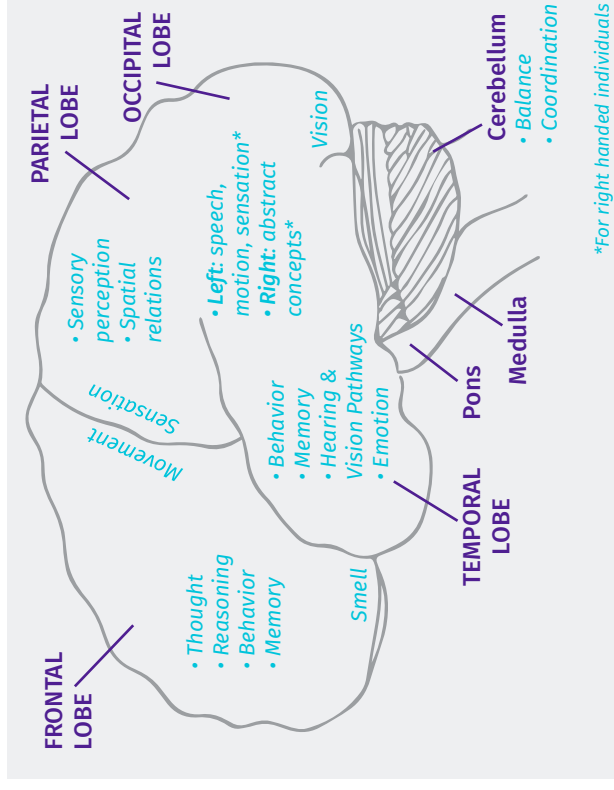
Email: _____

WHEN IS MY NEXT APPOINTMENT? WITH WHOM?

IMAGES:

(It is recommended that you keep a recent CD-ROM of your images and your reports with you, especially when you go to see doctors and when you travel.)

WHERE IS MY TUMOR?



I TAKE THESE MEDICATIONS:

(You are encouraged to ensure you have sufficient supply of your medications at all times, especially when you travel.)

MY ALLERGIES:

Chapter 2: Parts of the Brain

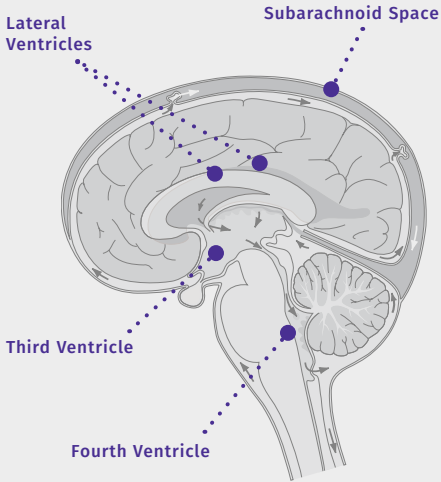
Groups of cells that are similar in appearance and perform the same function form tissue. The brain is a soft mass of supportive tissues and nerve cells connected to the spinal cord. Nerves in the brain and spinal cord transmit messages throughout the body.

The brain and spinal cord form the central nervous system (CNS). The central nervous system is the core of our existence. It controls our personality (*thoughts, memory, intelligence, speech, understanding and emotions*); our senses (*vision, hearing, taste, smell and touch*); our basic body functions (*breathing, heart beat and blood pressure*); and how we function in our environment (*movement, balance and coordination*).

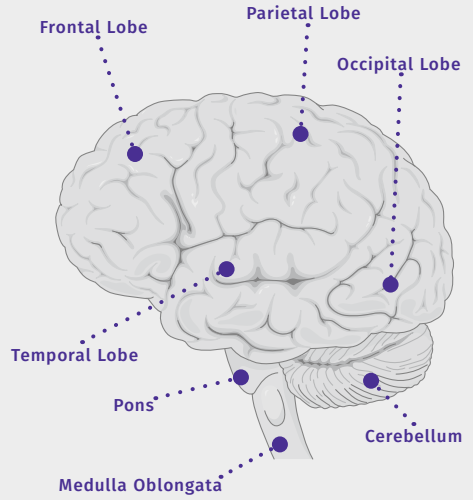
Learning about the normal workings of the brain and spine will help you understand the symptoms of brain tumors, how they are diagnosed and how they are treated.

> *Note: For a complete glossary of brain tumor terminology, see Chapter 12 “Brain Tumor Terminology”*

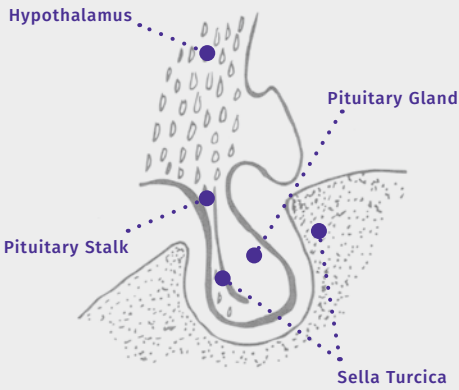
CSF AND VENTRICLES



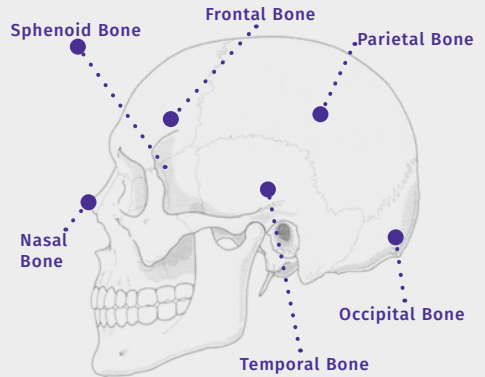
MAJOR PARTS OF THE BRAIN



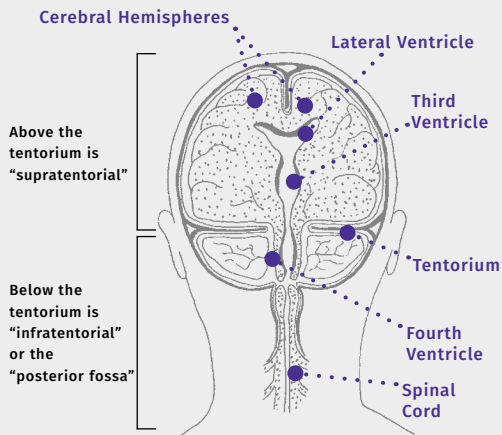
THE PITUITARY GLAND



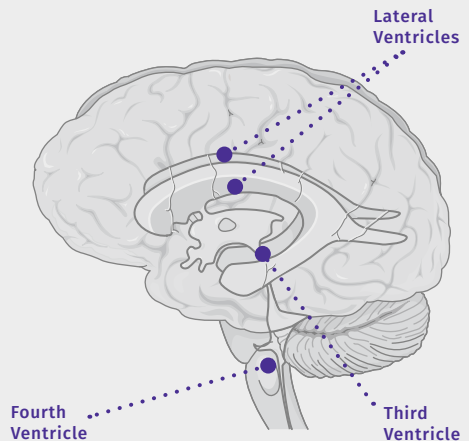
BONES OF THE SKULL



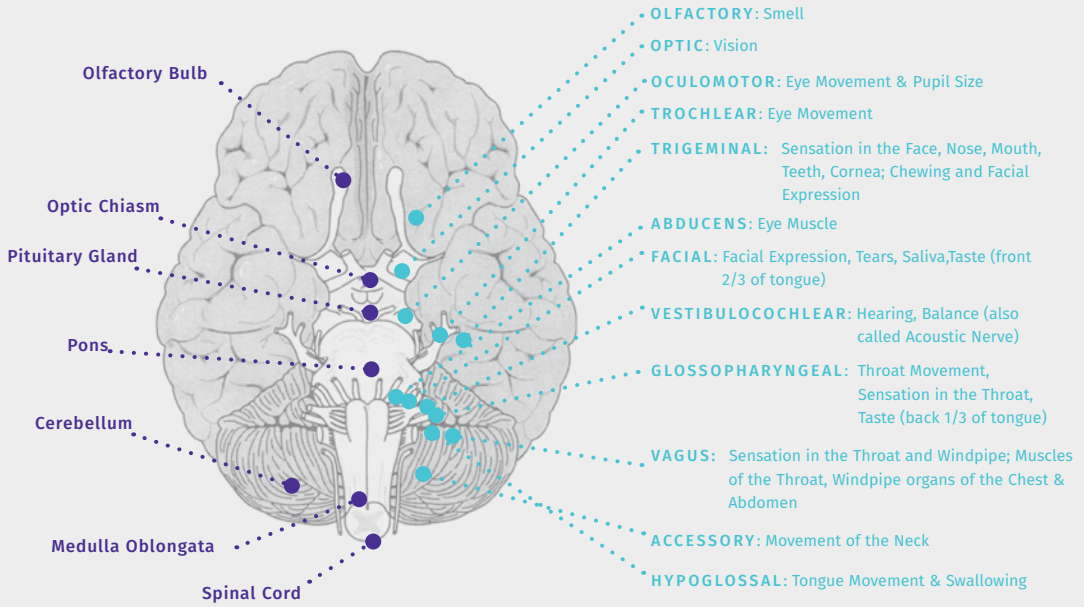
THE TENTORIUM



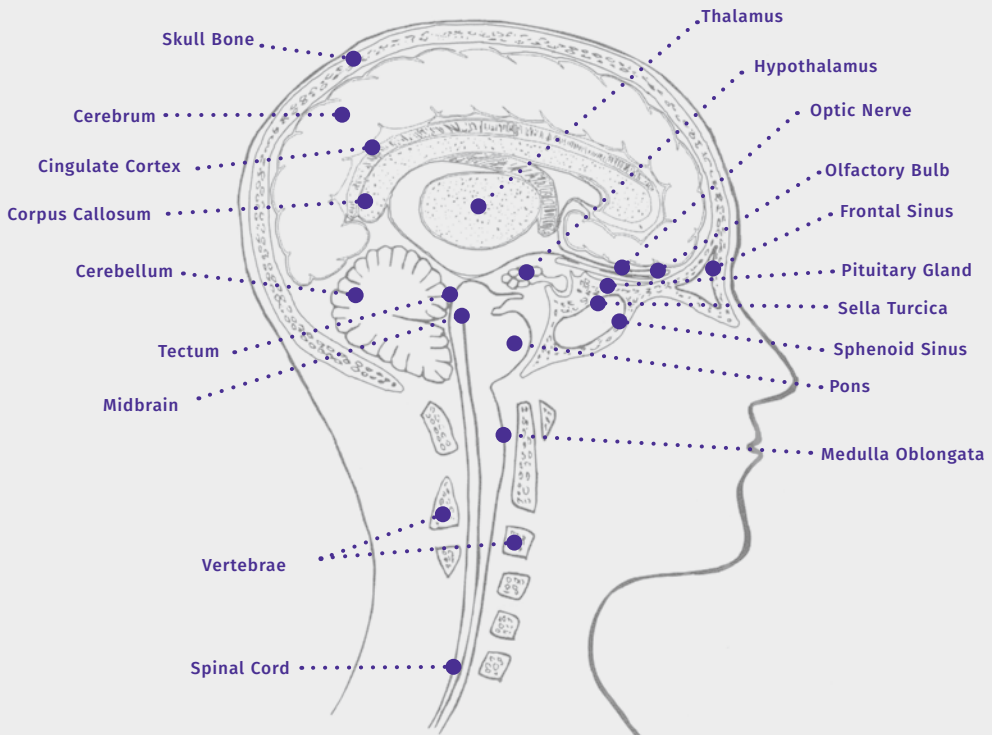
THE VENTRICLES



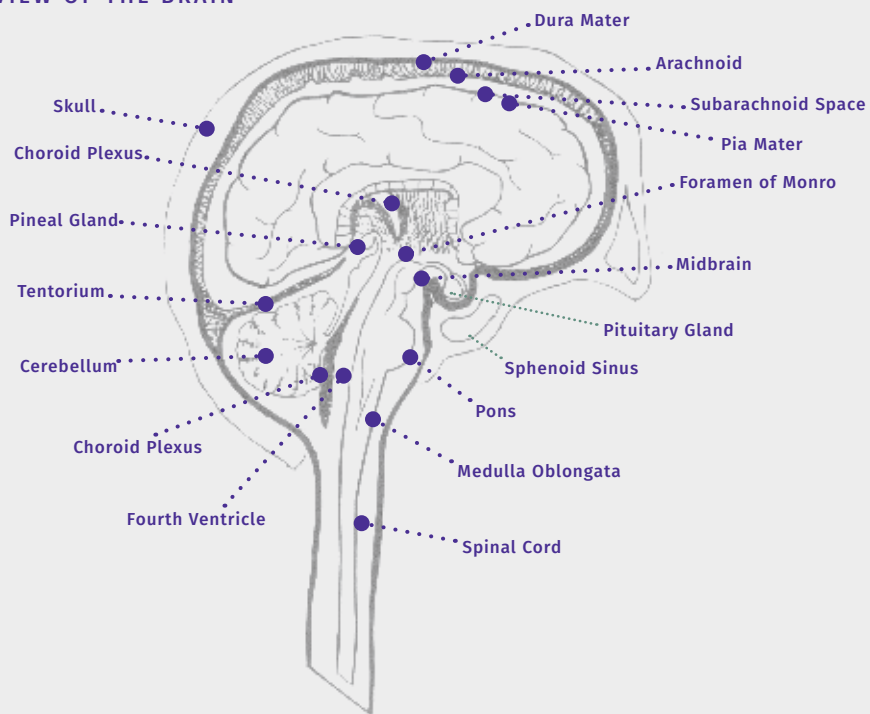
CRANIAL NERVES VIEW FROM BOTTOM OF BRAIN



CROSS SECTION OF THE BRAIN



SIDE VIEW OF THE BRAIN



Chapter 3: Types of Brain Tumors

This is an introduction to the more common brain tumors, their typical symptoms and locations, and their potential treatment options. Please remember that your tumor is unique and might not conform to the “average” characteristics described. Furthermore, treatments for brain tumors are constantly evolving. Please talk to your doctor about creating an individualized treatment plan for you or your loved one’s tumor.

The tumor names and their organization in this chapter are based on the World Health Organization (WHO) brain tumor classification system. Advanced analyses have revealed additional tumor characteristics that are refining our understanding of tumors and their respective presentations, treatments and outcomes.

For more information about specific tumors and treatments visit **[abta.org](https://www.abta.org)**.

ASTROCYTOMA

Astrocytomas are tumors that are thought to arise from astrocytes – cells that make up the “glue-like” or supportive tissue of the brain. These tumors are “graded” by the pathologist to indicate how normal or how abnormal the cells of the tumor look under the microscope. The WHO system grades astrocytomas on a scale from I to IV. Grade I tumors are usually localized (limited in growth) tumors that are often successfully treated through surgical removal. Grade II to IV tumors have increasing degrees of malignancy. Although surgery is beneficial, it does not completely cure these tumors. Grade II astrocytomas have slightly unusual looking cells. The cells of a grade III and IV astrocytoma are very abnormal in appearance. In this section we describe only the more common of these tumors: pilocytic astrocytoma, diffuse astrocytoma and anaplastic astrocytoma.

Pilocytic Astrocytoma

Also called *juvenile pilocytic astrocytoma*

These grade I astrocytomas are usually well-defined, non-infiltrating tumors – meaning they tend to stay in the area in which they started and do not spread into surrounding tissue. They generally form cysts or may be enclosed within a cyst. Although these are usually slow-growing tumors, they can become very large.

These tumors represent about 5–6% of all gliomas and are the most common glioma in children. They are generally diagnosed in children and young adults under the age of 20 and are rarely seen in older adults. The most common locations include the optic nerve (*optic glioma*), the optic chiasm near the hypothalamus, thalamus, basal ganglia, cerebral hemispheres and the cerebellum (*cerebellar astrocytoma*).

This tumor is the “most non-malignant” tumor of the astrocytomas. Pilocytic astrocytomas are generally considered benign tumors and are often cured by surgery alone. In adults and older children, radiation therapy might follow surgery if the tumor cannot be completely removed. Or, the residual (or left over) tumor may be carefully watched. In a “watchful waiting” situation, follow-up MRI scans are done at regular intervals to monitor for possible regrowth. If the tumor recurs, re-operation and some form of radiation are options. Some pilocytic tumors, such as most optic gliomas, cannot be safely removed because of their location and initial treatment may involve observation only.

The term *anaplastic (or malignant) pilocytic astrocytoma* is used only when the tumor has developed an extensive blood supply around the tumor or the tumor contains dead cells (necrosis). These rare tumors require more aggressive treatment than a non-malignant pilocytic astrocytoma.

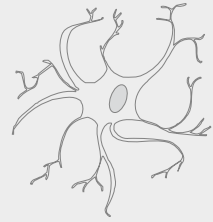
Diffuse astrocytoma

These grade II tumors have abnormal “tumor” cells mixed up with “normal” brain cells. Because of this, finding the border between where abnormal cells end and normal brain begin is impossible.

The overarching goal is for maximal safe resection. After surgery, most patients are at least considered for regional radiation, although this remains a very individualized decision between the patient and doctor. In an increasing number of cases, earlier use of chemotherapy is demonstrating a slower rate of tumor progression and an increased overall survival. Temozolomide is the most common chemotherapy used. Clinical trials, based on advanced analyses of your tumor, can also be considered as treatment options.

ASTROCYTE

Astrocytes are the cells that make up the “glue-like” or supportive tissue of the brain.



Anaplastic Astrocytoma

Also called grade III astrocytoma or malignant astrocytoma

An anaplastic astrocytoma is a grade III tumor. The word *anaplastic* means malignant. Astrocytomas often contain a mix of different cells and cells that are different grades, but brain tumors are graded by the highest grade (most abnormal) cell seen in the tumor. Even if a tumor is made of mostly grade II cells but has a few grade III cells, it will be given a grade of III. These tumors tend to have tentacle-like projections that grow into surrounding tissue.

This grade of tumor tends to occur in males more often than females and most frequently in people ages 45 and older.

The treatment options your doctor outlines will be based on the size and location of the tumor; what it looks like under the microscope; if and how far the tumor has spread; any previous treatment; and your general health. Generally, the first step in the treatment of an anaplastic astrocytoma is surgery. The goals of surgery are to obtain tumor tissue for diagnosis and treatment planning, to remove as much tumor as possible, and to reduce the symptoms caused by the presence of the tumor. There are some circumstances – such as certain medical conditions or concerns about the location of the tumor – in which a biopsy may be done in place of surgery. The tissue obtained during the biopsy is then used to confirm the diagnosis.

Because the tentacle-like cells of an astrocytoma grow into surrounding tissue, these tumors cannot be totally removed during surgery. Partial removal can help decrease symptoms and the tissue obtained during that surgery will confirm the type of tumor.

Radiation is then used to treat the remaining tumor. In general, the standard approach is external beam radiation directed to the area of tumor and a small area around it. Specialized delivery, such as the use of conformal radiation or intensity modulated radiation (IMRT) may be recommended. Although not standard treatment, there are other forms of radiation therapy available – focused or stereotactic radiosurgery and proton therapy – that may be recommended to you. Your radiation oncologist will decide which form of radiation therapy is best for your particular tumor.

Recently published international studies, with long-term follow-up, reveal that adding chemotherapy to radiation tends to lengthen the time of tumor control and also improve survival. Your doctor can discuss the specifics of chemotherapy with you or your loved one. Temozolomide is the most common chemotherapy drug used, but some treatment plans may still use the drugs BCNU, CCNU, procarbazine or cisplatin. Clinical trials, based on advanced analyses of your tumor, can also be considered as treatment options.

Anaplastic astrocytomas tend to recur and when they do, they may re-grow as a grade III or a grade IV tumor. Treatment is based on the grade of tumor at recurrence and location. For information on astrocytoma grade IV, see the section on glioblastoma.

BRAIN STEM GLIOMA

Brain stem gliomas arise in or on the brain stem – the area containing all of the intertwining connections from the brain to the spinal cord, as well as important structures involved in eye movements, face and throat muscle control, breathing and heart rate, and sensation.

Between 10-20% of brain tumors in children are brain stem gliomas. This tumor most often affects children between 5 and 10 years old, but can also be found in adults generally between 30 and 40 years old. Most of these tumors are astrocytomas, which vary from localized grade I tumors (mostly in children) to infiltrating grade II or III tumors. However, many are never biopsied due to the high-risk of performing any surgical procedure in that area, which makes determining the tumor grade impossible. In these situations, the diagnosis can usually be based from MRI scans.

Most of these tumors are classified by their location:

- > **Midbrain or tectum:** upper brain stem
- > **Pons:** middle brain stem
- > **Cervico-medullary:** lower brain stem

And by MRI appearance:

- > **Localized:** circumscribed or in one contained location
- > **Diffusely infiltrating:** tumor spread within the area
- > **Exophytic:** meaning the tumor has a knob protruding outside of the brainstem

The majority of brain stem tumors occur in the pons and are diffusely infiltrating, therefore preventing the tumor's surgical removal. A few of these tumors are localized and may be reachable for resection.

The symptoms of a brain stem glioma depend on the location of the tumor. The most common symptoms are related to eye movement abnormalities, which may cause double vision. Other symptoms include weakness or sensation changes of the face, swallowing difficulty, and hoarseness. Weakness, loss/changes in sensation or poor coordination on one side of the body may also occur. The tumor may also block the cerebrospinal fluid circulation resulting in hydrocephalus (dilatation of the fluid cavities in the brain), which can cause headaches, nausea, vomiting and unsteadiness in walking.

Treatment of a brain stem glioma is determined by the tumor location, grade and symptoms. Surgery may be feasible if a tumor appears localized or exophytic. The goals of surgery are to determine the grade and type of tumor and, in some cases, to remove the tumor. A shunt may also be placed if there is blockage of the cerebrospinal fluid circulation.

Radiation therapy may be used early if there are significant symptoms, or it may be postponed until the tumor grows or causes symptoms. Chemotherapy is used at diagnosis or if the tumor progresses following radiation therapy. The treatment plan is often based on whether imaging scans reveal characteristics similar to a grade II or a grade IV tumor. If the tumor appears to be a grade IV tumor, treatment similar to that used to treat glioblastoma (see glioblastoma section below) may be considered.

Radiation therapy with hyperfractionation (where the total dose of radiation is divided into small doses and treatments are given more than once a day) has been used in children in order to increase the effectiveness of the therapy and decrease side effects. Unfortunately, this has not resulted in significant advantage over standard radiation.

CRANIOPHARYNGIOMA

This is a non-malignant tumor arising from small nests of cells located near the pituitary stalk. Craniopharyngiomas represent 2–5% of all primary brain tumors and 5–10% of childhood brain tumors. There are two age groups in which this tumor tends to be seen: those up to age 14 and again after age 45.

Adamantinomatous (ordinary) craniopharyngioma occurs in children and tends to be more cystic than the papillary craniopharyngioma. The papillary craniopharyngioma occurs in adults and is a more solid tumor.

Craniopharyngiomas occur in the sellar region, near the pituitary gland. They often involve the third ventricle, optic nerve and pituitary gland. These localized tumors may reach a large size before they are diagnosed.

Most symptoms associated with this tumor are a result of increased intracranial pressure due to obstruction of the foramen of monro (one of the small tunnels through which cerebrospinal fluid exits the ventricles). Other symptoms result from pressure on the optic tract and pituitary gland. Obesity, delayed development, impaired vision and a swollen optic nerve can also be common symptoms.

Surgery to remove the tumor is usually the first step in treatment. If hydrocephalus (brain swelling) is present, a shunt may be placed during surgery. That shunt will help drain excess cerebrospinal fluid away from the brain. A form of radiation therapy may be suggested if all of the visible tumor cannot be removed. This may include a focused form of radiation, such as radiosurgery or conformal radiation. In children younger than age 3, radiation therapy may be delayed by the use of surgery or hormone therapies. Because this tumor tends to be located close to the pituitary gland – which controls hormone balance in the body – an endocrinologist may become involved in the long-term care plan. An endocrinologist is a doctor trained in treating hormone imbalances.

EPENDYMOMA

Ependymomas may originate from ependymal cells (which line the ventricles of the brain and the center of the spinal cord) or from radial glial cells (cells related to early development of the brain). These are relatively rare tumors, accounting for less than 2% of all primary tumors and about 7% of all gliomas. They represent about 5% of childhood brain tumors.

Ependymomas are soft, greyish or red tumors which may contain cysts or mineral calcifications. They are divided into four major types:

- > **Subependymomas** (grade I) are slow growing and usually occur near a ventricle.
- > **Myxopapillary ependymomas** (grade II) are slow growing and tend to occur in the lower part of the spinal column.
- > **Ependymomas** (grade II) are usually located along, within or adjacent to the ventricular system; often in the posterior fossa or in the spinal cord. Based on the appearance of the cell patterns when viewed under a microscope, this group of tumors can be sub-divided into smaller groups based on the appearance of their cell patterns: cellular ependymomas, papillary ependymoma, clear cell ependymoma and tancytic ependymoma. There are several other patterns as well, but regardless of appearance, these are all considered grade II tumors.
- > **Anaplastic ependymomas** (grade III) tend to be faster growing tumors. These are most commonly found in the brain in adults, and specifically in the posterior fossa in children. They are rarely found in the spinal cord.

EPENDYMOMA

Ependymal cells line the ventricles of the brain and the center of the spinal cord.



The first step in the treatment of an ependymoma is surgery to remove as much tumor as possible. The amount of tumor that can be removed, however, depends on the location of the tumor. Radiation therapy is usually recommended for older children and adults if all visible tumor was not removed during surgery. In some cases, radiation therapy is used even after complete resection. If the tumor is localized (contained to one area), radiation therapy is usually given just to that area of the brain. If the tumor has spread, radiation is usually given to the entire brain and spine, with an extra amount of radiation (called a “boost”) given to the area of the brain where the tumor started.

In general, the role of chemotherapy in treating newly diagnosed ependymomas is not clear. However, chemotherapy may be used to treat tumors that have grown back after radiation therapy or to delay radiation therapy in infants and young children. Clinical trials, if available, can also be considered as a treatment option.

GERM CELL TUMORS

These uncommon tumors represent less than 4% of brain tumors in children and adolescents (ages 0-19) and occur primarily in young people between the ages of 11 and 30. Germ cell tumors arise in the pineal or suprasellar regions of the brain. Included in this type of tumor are the germinoma, the teratoma, the more aggressive embryonal carcinoma and yolk sac (endodermal sinus) tumors, and the choriocarcinoma. Mixed germ cell tumors also exist. Because all of these tumors tend to spread via the cerebrospinal fluid (CSF), diagnosis includes evaluation of the entire brain and spinal cord. An MRI scan and examination of the CSF for the presence of tumor cells is used for that evaluation.

Germ cell tumors are the only primary brain tumors that might be diagnosed by tumor markers found in the cerebrospinal fluid and blood. The markers are alpha-fetoprotein (AFP), placental alkaline phosphatase (PAP) and human chorionic gonadotropin (HCG). More commonly, however, the markers are used to monitor the effectiveness of therapy and to monitor signs of recurrence.

Because of their location, most germ cell tumors are treated with chemotherapy or a combination of radiation and chemotherapy, rather than surgery. However, a biopsy to establish an exact diagnosis is not uncommon, and some very experienced surgeons have had success removing certain pineal region tumors. Surgery may be required to treat hydrocephalus (buildup of fluid in the brain) caused by the tumor blocking the cerebrospinal fluid pathways.

GLIOBLASTOMA

Also called “astrocytoma, grade IV” and “GBM”

Glioblastomas are malignant Grade IV tumors, where a large portion of tumor cells are reproducing and dividing at any given time. They are nourished by an ample and abnormal tumor vessel blood supply. The tumor is predominantly made up of abnormal astrocytic cells, but also contain a mix of different cell types (including blood vessels) and areas of dead cells (necrosis). Glioblastomas are infiltrative and invade into nearby regions of the brain. They can also sometimes spread to the opposite side of the brain through connection fibers (corpus callosum). It is exceedingly rare for glioblastomas to spread outside of the brain.

Glioblastomas may arise de novo, meaning they begin as a Grade IV tumor with no evidence of a lower grade precursor. De novo tumors are the most common form of glioblastoma and tend to be more aggressive and tend to affect older patients. Alternatively, secondary glioblastomas may progress from a lower-grade astrocytic tumors (Grade II or Grade III) and evolve into Grade IV tumors over time. In general, these tumors tend to be slower growing initially, but can progressively become aggressive.

Glioblastomas are usually diagnosed as either IDH-wildtype or IDH-mutant (please see the Genetic Profile section below for more information). IDH-wildtype glioblastomas are more common, tend to be more aggressive, and have worse prognosis than IDH-mutant glioblastomas.

Glioblastomas are generally found in the cerebral hemispheres of the brain, but can be found anywhere in the brain. IDH mutant glioblastomas tend to arise preferentially in the frontal lobe.

Patients with glioblastomas develop symptoms rapidly due to mass effect from the tumor itself or from the fluid surrounding the tumor (edema) that causes further brain swelling. For example, common symptoms at diagnosis are related to the increased pressure in the brain (nausea, vomiting, and severe headaches which are typically worse in the morning). Patients can also present with neurological symptoms which are dependent on the tumor location (for example,

weakness or sensory changes of face, arm or leg, balance difficulties and neurocognitive/memory issues). Other common presentation includes seizures.

Glioblastoma can be difficult to treat since some cells may respond well to certain therapies, while others may not be affected at all. Because of this, the treatment plan for glioblastoma may combine several approaches.

The first step in treating glioblastoma is a surgical procedure to make a diagnosis, to relieve pressure on the brain, and to safely remove as much tumor as possible. Glioblastomas are diffuse and have finger-like tentacles that infiltrate the brain, which makes them very difficult to remove completely. This is particularly true when the tumors are growing near important regions of the brain that control functions such as language and movement/coordination.

Radiation and chemotherapy are used to slow down the growth of residual tumor after surgery and for tumors that cannot be removed with surgery. Additional treatments such as angiogenesis inhibitors may be used for tumors that recur or those that are non-responsive as a second-line agent. Tumor Treating Fields (TTFields) may be also be offered especially for recurrent tumors in adults.

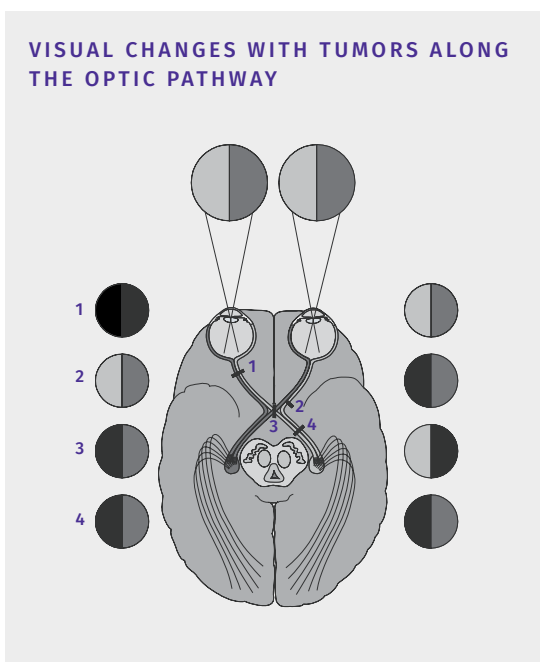
With standard treatment, median survival for adults with glioblastoma, IDH-wildtype, is approximately 11-15 months.

There are factors that can contribute to improved prognosis, such as younger age at diagnosis (less than 50 years), near-complete removal of the tumor in surgery. Important molecular markers are determined after biopsy or surgery, which provide information for diagnosis and prognosis. For patients with IDH mutant glioblastoma, the prognosis is significantly better (median survival of 27 – 31 months) compared to IDH wildtype glioblastoma (median survival 11-13 months) after diagnosis. Another marker, methylation of a gene called MGMT promoter is also an important marker. MGMT is important for the stability of genes in all cells. When it is methylated, it is inactivated. This makes cancer cells more sensitive to certain chemotherapy drugs such as temozolomide because the DNA gets so damaged that the cells die.

Glioblastomas represent about 15% of all primary brain tumors. Glioblastomas are slightly more common in men than in women. IDH mutant glioblastomas account for approximately 10% of all glioblastomas.

Clinical Trials

A clinical trial (also called an investigational study) offers new or experimental treatments to qualified brain tumor patients. Clinical trials are tests to determine if a particular treatment is safe and effective for use. Clinical trial participants volunteer to receive a treatment that otherwise would not be available to them.



GLIOMA

This is a general term for any tumor that arises from the supportive glia tissue of the brain. This tissue helps to keep the neurons (“thinking cells”) in place and functioning well. There are three types of normal glial cells that can give rise to tumors.

- > **Astrocytes** are star-shaped cells that can give rise to astrocytomas (including glioblastomas)
- > **Oligodendrocytes** are cells with short arms forming the insulation of neurons that can give rise to oligodendrogliomas
- > **Ependymal cells** form the lining of the fluid cavities in the brain and can give rise to ependymomas



Occasionally, tumors will display a mixture of these different cells and are called mixed gliomas.

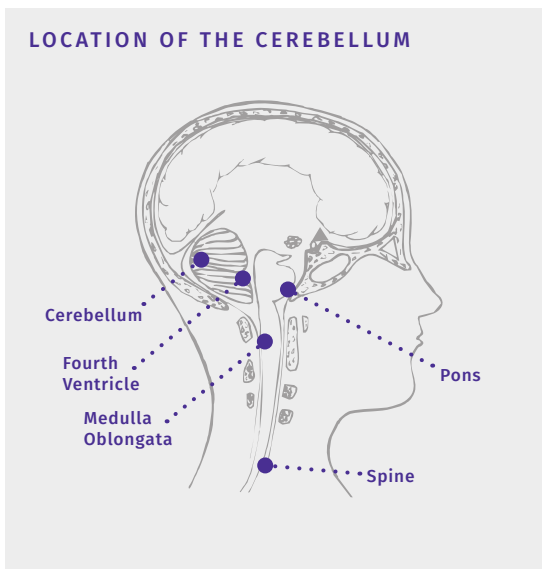
Names such as *optic nerve glioma* and *brain stem glioma* refer to the location of these tumors and not the type of tissue that gave rise to them. A specific diagnosis is only possible if a sample of the tumor is obtained during surgery or biopsy.

MEDULLOBLASTOMA

Medulloblastomas represent about 7% of the brain tumors in children and adolescents (ages 0-19). Medulloblastomas are always located in the cerebellum. There are five known subtypes of medulloblastoma based on advanced analyses and these subtypes correlate with presentation, availability of treatments and trials, response to specific treatments, and outcome.

Medulloblastoma is a fast growing, high-grade tumor that frequently spreads to other parts of the central nervous system. Given its location – close to one of the fluid cavities of the brain, called the fourth ventricle – the tumor may also extend into that cavity and block the cerebrospinal fluid circulation or send tumor cells through the spinal fluid to the spine. It is uncommon for medulloblastomas to spread outside the brain and spinal cord.

The most common symptoms of medulloblastoma, particularly in young children, include behavioral changes; symptoms of increased intracranial pressure such as headaches, nausea, vomiting and drowsiness; unbalanced walking and poor coordination of the limbs; and unusual eye movements.



At diagnosis (and during and after treatment) testing will also be done to check for possible tumor spread, including an MRI of the spine and a cerebrospinal fluid analysis.

Treatment consists of surgical removal of as much tumor as possible, radiation and chemotherapy. Optimal treatment in adolescents and adults is much less defined as in infants and children.

After surgery, radiation to the tumor area followed by a lower dose of radiation to the entire brain and spinal cord is typically used for older children, for adults that show no sign of the tumor spreading, and for patients

who had most of the tumor removed. Very young children are often treated with chemotherapy instead of radiation.

Chemotherapy generally follows radiation therapy. The most commonly used agents include a combination of cisplatin and vincristine with either cyclophosphamide or CCNU. Other drugs, such as etoposide, have also shown activity against the tumor.

There is no standard treatment for recurrent tumors. Some patients with a recurrent tumor who show good response to chemotherapy may benefit from high dose chemotherapy with autologous stem cell transplant.

Clinical trials can also be considered as a potential treatment option.

METASTATIC BRAIN TUMORS

The terms metastatic brain tumor, brain metastasis or secondary brain tumor refer to cancer that begins elsewhere in the body and spreads to the brain.

Metastatic brain tumors usually contain the same type of cancer cells found at the primary site. For example, small-cell lung cancer that moves to the brain forms small-cell cancer in the brain. Squamous-cell head and neck cancer forms squamous-cell cancer in the brain. Brain metastasis can present as a single tumor or multiple tumors.

The symptoms of metastatic brain tumors are the same as those of primary brain tumors, and are related to the location of the tumor within the brain. Each part of the brain controls specific body functions. Symptoms appear when areas of the brain can no longer function properly. Headache and seizures are the two most common symptoms. Disturbance in the way one thinks and processes thoughts (cognition) is another common symptom of a metastatic brain tumor. Cognitive challenges might include difficulty with memory (especially short term memory) or personality and behavior changes. Motor problems, such as weakness on one side of the body or an unbalanced walk, can be related to a tumor located in the part of the brain that controls these functions. Metastatic tumors in the spine may cause back pain, weakness, or changes in sensation in an arm or leg, or may cause loss of bladder/bowel control. Both cognitive and motor problems may also be caused by edema, or swelling, around the tumor.

If a brain scan shows a suspected brain tumor, your next step will likely be a consultation with a neurosurgeon, radiation oncologist or medical/neuro-oncologist. The neurosurgeon will look at your scans to determine if the tumor(s) can be surgically removed or if other treatment options would be more reasonable. The three main categories of treatments include surgery, radiation (whole brain radiation, stereotactic radiosurgery, or both) and medical therapy (chemotherapy, targeted therapy or immune-based therapy). More than one type of treatment might be suggested.

When planning your treatment, your doctor will take several factors into consideration:

- > Your history of cancer
- > The status of that cancer
- > Your overall health
- > Number and size of metastatic tumors
- > Location of the metastatic tumor(s) within the brain or spine

Early treatment of your brain tumor will focus on controlling symptoms, such as swelling of the brain and/or seizures. If you have a limited number of metastatic brain tumors (generally one to three tumors, or a small number of tumors that are close to each other) and if the primary cancer is treatable and under control, your treatment plan may include surgery to confirm the diagnosis

and remove the tumor, followed by a form of radiation therapy. This is generally followed by medical therapy that may impact not only the primary cancer but also metastatic brain tumor.

If you have multiple brain metastases (four or more brain tumors), traditionally whole-brain radiation therapy has been recommended; however, recently the use of radiosurgery or medical therapy has increased. If there is a question about the scan results or the diagnosis, a biopsy or surgery to remove the brain tumors may be done. This will allow your physicians to confirm that the brain tumors are related to the primary cancer. Metastases to the spine may be treated with radiation therapy alone or with surgery plus radiation.

Research shows that the number of metastases is not the sole predictor of how well you might do following treatment. Your neurological function (how you are affected by your brain metastases), the status of the primary cancer site (i.e. the presence/absence of metastases in other parts of the body), type of cancer and the genetic alterations in the cancer also appear to influence overall survival. Treatment decisions will take into account not only long term survival possibilities, but your quality of life during and after treatment, as well as cognition concerns.

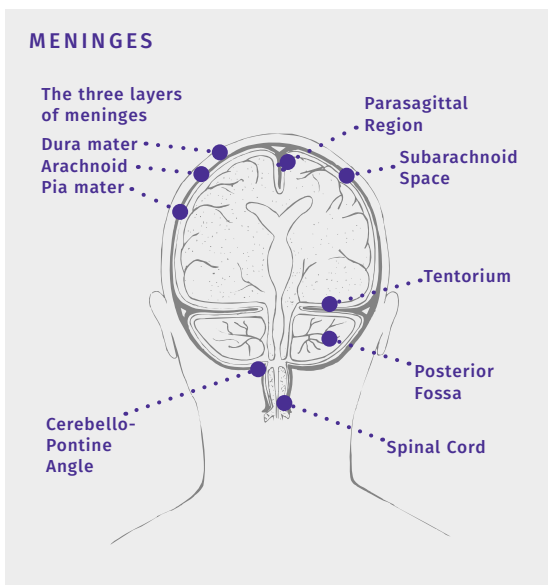
Metastatic brain tumors are the most common brain tumor in adults. The exact incidence of metastatic brain tumors is not known but is estimated between 200,000 and 300,000 people per year.

MENINGIOMA

These tumors arise from the “arachnoid mater” – one of the layers of the meninges (the lining of the brain). Meningiomas represent about 37% of all primary brain tumors and occur most frequently in middle-aged women. The majority of meningiomas are non-malignant, grade I, slow-growing tumors that are localized and non-infiltrating.

Meningiomas are most often located:

- > Between the cerebral hemispheres (called *parasagittal meningiomas*)
- > Within the protective tissues that cover the spinal cord and brain, known as the meninges (called *convexity meningiomas*)
- > At the base of the skull
- > In the back, lower part of the brain called the posterior fossa.



They occur less frequently in the spine. Most often a single tumor is found, but multiple meningiomas also occur.

Risk factors for meningioma include prior radiation exposure to the head and a genetic disorder called “neurofibromatosis type 2” (read more in Chapter 4, under “Genetic Factors”), which affects the nervous system and the skin; however, meningiomas also occur in people who have no risk factors.

A variety of symptoms can occur, depending on the tumor’s location. The most common indications are headaches, weakness on one side, seizures, personality and behavioral changes, and confusion. Neuro-imaging (scanning) with a CT or MRI is used to evaluate the location of the tumor. Calcifications may be seen in cases of slow growing meningioma.

Non-malignant meningiomas (grade I) are slow-growing with distinct borders. Because they grow slowly, they can grow quite large before symptoms become noticeable. Symptoms are caused by compression rather than by the tumor growing into brain tissue.

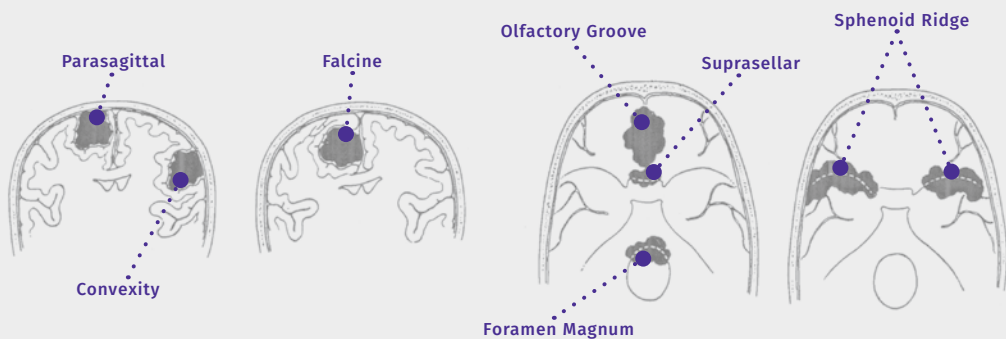
If the tumor is accessible, the standard treatment for meningiomas is surgery. During the procedure, the surgeon will try to remove the tumor, the portion of the dura mater (the outermost layer of the meninges) that the tumor is attached to, and any bone that is involved. Total removal appears critical for long-term tumor control.

Evaluation of the blood supply of the tumor may be done before surgery, and in some cases the blood vessels are embolized (purposefully blocked) to facilitate the removal of the tumor.

If the tumor is not entirely removed, radiation therapy or radiosurgery may occur after surgery. For some patients, surgery may not be recommended. Long-term, close observation with scans is typically recommended for patients that have no symptoms (usually meaning they have been diagnosed coincidentally), for patients with minor symptoms of long duration, and for patients that cannot receive surgery due to the location of the tumor. An alternative includes focused radiation, also called “stereotactic radiosurgery.”

Atypical meningiomas (grade II) have a middle range of behavior. These tumors are not clearly

COMMON LOCATIONS OF MENINGIOMAS



Burger, Scheithauer and Vogel. *Surgical Pathology of the Nervous System and Its Coverings*. Fourth Edition. Churchill Livingstone, New York, 2002. Diagram reproduced with permission.

malignant, but they may invade the brain, have a tendency to recur and are faster-growing. The diagnosis and grade are determined by specific features that can be seen under the microscope. Radiation therapy is indicated after surgery, particularly if any residual tumor is present.

Anaplastic or malignant meningiomas (grade III) and *papillary meningiomas* are malignant and tend to invade adjacent brain tissue. They represent less than 5% of meningiomas. Radiation therapy is clearly indicated following surgery regardless of whether residual tumor is present.

Meningiomas may recur, either as a slow-growing tumor or sometimes as a more rapid growing, higher-grade tumor. Recurrent tumors are treated similarly, with surgery followed by either standard radiation therapy or radiosurgery, regardless of the grade of the meningioma. Chemotherapy and biological agents are being studied for recurrent meningioma. Hormone therapy does not appear effective. Advanced analyses of meningiomas is leading to potential medicines, and clinical trials, if available, can also be considered as a treatment option.

MIXED GLIOMA/OLIGOASTROCYTOMA

Oligoastrocytomas were thought to be mixed glioma tumors, containing both abnormal oligodendroglioma and astrocytoma cells. In the 2016 World Health Organization classification, the diagnosis of Oligoastrocytoma is strongly discouraged. Nearly all tumors with features suggesting both cell types can be classified as either Astrocytoma or Oligodendroglioma using genetic testing. If they exist, true Oligoastrocytomas are very rare. The behavior of a mixed glioma tumor tends to be based on the grade of the tumor. The tumor behavior may reflect the activity of the most abundant cell type.

Standard treatment for a mixed glioma is similar to that for an astrocytoma and oligodendroglioma of the same grade. The treatment plan may include surgery followed by radiation therapy, particularly if the tumor is high-grade (grade III or IV). Although, it may also be indicated for lower-grade tumors (grade II). Chemotherapy will generally be used in high-grade tumors.

OLIGODENDROGLIOMA

These tumors arise from oligodendrocytes, one of the types of cells that make up the supportive, or glial, tissue of the brain. Under the microscope, these tumor cells seem to have a fried-egg shape with “short arms” as opposed to astrocytomas, which are star-like shape with “long arms.” The most recent WHO guidelines have determined these tumors by advanced analyses, specifically the co-deletion of 1p/19q and IDH-mutation.

Oligodendrogliomas can be low-grade (grade II) or high-grade (grade III, also called anaplastic). Sometimes oligodendrogliomas may be mixed with other cell types. The grade denotes the speed with which the tumor cells reproduce and the aggressiveness of the tumor.

Oligodendrogliomas occur most frequently in young and middle-aged adults, but can also be found in children. They are most commonly located in the cerebral hemisphere, with about half of those tumors being found in the frontal lobe. Seizures are the most common initial symptom, particularly in low-grade tumors.

Surgical removal is the standard treatment for accessible tumors. Biopsy alone may be performed for inaccessible tumors. The tumor sample removed during a biopsy is used to confirm the diagnosis and the grade of tumor.

For low-grade oligodendroglioma that appear on the MRI scan after surgery to have been completely resected, close observation with follow-up MRIs may be recommended. If some of the tumor remains after surgery (this is called “residual” tumor), radiation therapy and chemotherapy may be recommended in some patients. However, the individual patients who could benefit from these treatments, and the best timing to conduct these treatments (immediately or at tumor progression) is being determined in clinical trials.

Available results from international studies with long-term follow-up reveal that the addition of chemotherapy to radiation results in both maintaining tumor control and improved survival. Recurrent low-grade oligodendrogliomas can be treated with surgery, radiation therapy (if not given initially) or chemotherapy.

For anaplastic oligodendroglioma, a combination of radiation therapy and chemotherapy, such as PCV (procarbazine, CCNU and vincristine) or temozolomide, is suggested. Recurrent anaplastic oligodendroglioma may be treated with surgery and/or chemotherapy.

OLIGODENDROCYTE

Oligodendrocytes are one of the types of cells that make up the supportive, or glial, tissue of the brain.



As introduced above, advanced analyses of oligodendroglioma have shown that combined loss of the short arm of chromosome 1 and the long arm of chromosome 19 (called “1p 19q loss”) is associated with improved outcome. These tumors are also known for IDH-mutation, a mutation in an enzyme of tumor metabolism. Clinical trials are available for newly diagnosed and recurrent low-grade or high-grade oligodendrogliomas. Many of these trials take into account the genetic features of the tumor, thereby highlighting the importance of obtaining tumor tissue for analysis.

RECURRENT TUMORS

Many tumors cannot be removed completely during surgery because they have invaded the surrounding normal tissues. Some tumors – such as low-grade gliomas (astrocytomas and oligodendrogliomas) and meningiomas – have the potential to recur as higher-grade or more aggressive tumors. If the tumor recurs, a second surgery may be indicated. Conventional radiation therapy can be given if it was not used initially. A form of focused radiation therapy, such as stereotactic radiosurgery, might be recommended if conventional radiation therapy has already been given. Chemotherapy is frequently used to treat recurrent tumors. Clinical trials with chemotherapy and biologic therapies are available for recurrent high-grade gliomas and can be considered as another treatment option.

ADDITIONAL TUMOR TYPES

More information on brain tumor types, treatments, clinical trials and other brain tumor resources can be found at www.abta.org or by calling 800-886-ABTA (2282).

- > Acoustic Neuroma
- > Atypical Teroid Rhabdoid Tumor (ATRT)
- > Chondroma, Chondrosarcoma, Chordoma
- > Choroid plexis tumors
- > Cysts
- > Dysmbryoplastic neuroepithelial tumor (DNT)
- > Gangliocytoma
- > Ganglioglioma
- > Germinoma
- > Glioma, optic
- > Gliomatosis cerebri
- > Glomus jugulare
- > Hemangioblastoma
- > Hemangipericytoma
- > Lipoma
- > Neuroblastoma cerebral
- > Neurocytoma central
- > Neurofibromatosis
- > Pinel
- > Pituitary
- > PNET
- > Pseudotumor cerebri
- > Schwannoma
- > Skull base
- > Spinal cord
- > Teratoma
- > Tuberos sclerosis
- > Vestibular schwannoma

Chapter 4: Causes and Risk Factors

When patients learn they have an uncommon disease, questions may arise about the causes and risks for that disease.

- > *Why did this happen to me?*
- > *What do I have in common with other people who have this disease?*
- > *What does this mean for my family?*
- > *How close are we to preventing this?*

These are all normal questions.

An epidemiologist is a scientist trained in studying groups of people with the same disease. Brain tumor epidemiologists look for causes and risk factors that help to explain why people develop brain tumors and what these people have in common with each other. These observations of “commonality” can provide important clues that can show links between individuals. If an epidemiologist believes he or she has discovered a link, it must be replicated in additional studies conducted by other scientists (a process called validation). If the finding is successfully replicated, then it can be considered a convincing cause or risk factor for that disease.

INTRODUCTION

Causes and risk factors can be either environmental or genetic. Environmental factors are something you are exposed to in your surroundings, whereas genetic factors are inherited from your parents.

Unfortunately, no risk factor has been identified that accounts for the majority of brain tumors, even though many environmental and genetic factors have been, and are currently being studied.

ENVIRONMENTAL FACTORS

Many studies have looked at a wide spectrum of environmental factors as possible causes of brain tumors, including but not limited to:

- > Being exposed to air pollution, residential power lines, secondhand smoke, agricultural chemicals and industrial formaldehyde
- > Using cell phones
- > Working in synthetic rubber manufacturing or petroleum refining/production
- > Smoking cigarettes, smoking cigarettes while pregnant and consuming alcohol
- > Using common medications like birth control pills, sleeping pills, headache remedies, over-the-counter pain treatments and allergy-relief medications
- > Having a history of head trauma, epilepsy, seizures or convulsions
- > Experiencing viruses and common infections
- > Consuming cured foods (nitrites)

These exposures are difficult to accurately measure and can lead to inconsistencies across studies, making the results difficult to validate. Additional long-term research on these factors is needed before definite conclusions can be formed.

Of the long list of factors studied, only exposure to ionizing radiation has been consistently associated with an increased risk for developing a brain tumor. Ionizing radiation uses “high-frequency” energy waves, such as X-rays or gamma rays. However, radiation doses used today for medical and dental therapies are better focused than those used in medicine decades ago.

On the other hand, some studies have shown that a history of allergies as an adult, eating fruits and vegetables as a child and having a mother who ate fruits and vegetables during pregnancy, and having chicken pox as a child puts one at a decreased risk of developing a brain tumor.

Over the last decade, the association between using a cell phone and developing a brain tumor has been of particular interest. Multiple large studies have been performed in both the United States and Europe. Some have shown an association between cell phone use and brain tumor risk, while other studies show no association. In addition, studies have also investigated the difference in risk of a brain tumor between short-term and long-term (more than 10 years) cell phone use, but these studies have also produced conflicting results. In general, the conclusions from most of these studies are:

- > There is no consistent association between cell phone use and risk of developing a brain tumor (malignant or non-malignant)
- > There may be a very slight increased risk of a brain tumor associated with using a cell phone for 10 years or more

Multiple large studies have been performed to see if brain tumors are occurring more frequently now that cell phone use is commonplace. If cell phones were a major cause of brain tumors, the expectation would be that the frequency of brain tumors would greatly increase; however, these studies have largely found that there have been no large changes in the frequency of brain tumors over time. Further studies, in both the laboratory and in humans with longer follow up, are needed to fully understand this exposure and any potential relationship it has with brain tumor development.

Ultimately, additional research is needed before definite conclusions can be formed.

ABOUT CLUSTERS OF BRAIN TUMORS

Understandably, communities become concerned when several individuals within a neighborhood are diagnosed with brain tumors. Scientists studying these groups will want to learn whether these are metastatic brain tumors (those that began as cancer elsewhere in the body and moved to the brain) or primary brain tumors (those that began in the brain or spinal cord and tend to stay there). If the brain tumors are primary tumors, scientists will want to know the specific type(s). Clusters involving the same type of primary brain tumor are most concerning as these tumors may share similar biologic origins. Metastatic brain tumors – such as breast cancer, lung cancer or colon cancer that spread to the brain – most likely do not share the same origins as primary brain tumors.

The first step in reporting a perceived cluster of brain tumors is to call your local health department. They can tell you if the incidence of brain tumors is higher than expected for the area or if any current investigations are underway. Once reported to the local health department, the next level of authority may be the county or the state department of health. Each state hires epidemiologists to monitor the incidence of disease in their state. They also have the authority to order an investigation, if warranted.

GENETIC FACTORS

Genes are the operating instructions for the entire body. Anything that refers to our genes can be called *genetic*.

Genetic factors refer to conditions or diseases inherited within families. Only 5–10 percent of all cancer is actually hereditary (inherited from one generation to another in a family). There are a few rare, inherited genetic syndromes that involve brain tumors; thus, there are very few families where multiple family members have a brain tumor. In those syndromes, a mutation in a specific gene has to be passed from grandparent to parent to child. These syndromes, along with the inherited gene, are: Neurofibromatosis 1 (NF1 gene), Neurofibromatosis 2 (NF2 gene), Turcots (APC gene), Gorlins (PTCH gene), Tuberous Sclerosis (TSC1 and TSC2 genes) and Li-Fraumeni syndrome (TP53 gene). In addition to these known inherited genetic syndromes, there may be other inherited mutations within families that increase risk for a brain tumor. For example, one large study has identified inherited rare mutations (in families with brain tumors) in a group of genes that protect the ends of chromosomes, or telomeres, from damage.

> **Genetic** means anything referring to our genes.

> **Inherited or hereditary** means a disease or condition passed from one generation to another within a family.

With the publication of the Human Genome Project and advances in genotyping technology, scientists can now identify over a million genetic variants found in the human body and ask the question: “Are any of these inherited genetic variants associated with risk of a brain tumor?”

This type of study is called a genome-wide association (GWA) study. One recent GWA study of glioma found 25 commonly occurring inherited genetic variants that slightly increased risk for glioma. This research shows that common genetic differences within the general population can contribute to risk for developing a malignant brain tumor. Much more investigation is needed to fully understand the importance of these variations and how they may impact brain tumor risk. This type of GWA study has yet to be performed for non-malignant brain tumors or pediatric brain tumors.

The vast majority of genetic risk factors are not inherited at birth, but actually accumulate over time as we age (also called somatic or acquired). While most of our genes do their jobs as expected, a small number may become inactive or start functioning abnormally. The end result of having an abnormal gene can be as simple as two different colored eyes or as complex as the onset of a disease. There are many different types of genes that are thought to not be working correctly in brain tumors, some of which are targets of currently used therapies for cancer:

- > **Tumor suppressor** genes make proteins that stop tumor growth in normal cells. The most well-defined tumor suppressor gene is TP53, which is believed to play a role in causing a low-grade malignant brain tumor to develop into a high-grade malignant brain tumor.
- > **Oncogenes** make proteins that cause cells to grow in an out-of-control manner.
- > **Growth factors** play a role in making sure that cells grow normally. Epidermal Growth Factor Receptor (EGFR) is a growth factor that has been well studied in brain tumors because it has been seen in very high quantities in high-grade malignant brain tumors, causing these tumors to grow abnormally fast.
- > **Cyclin-dependent kinase inhibitors** play a role in making sure that the cell goes through its growth cycle normally.
- > **DNA repair genes** make proteins that control accurate repair of damaged DNA.
- > **Carcinogen metabolizing genes** make proteins that break down toxic chemicals in the body that could cause damage to one's DNA, like the chemicals in cigarette smoke and/or alcohol.
- > **Immune response genes** make proteins that control how one's immune system responds to viruses and infections.

Acquired or somatic means genetic changes that have accumulated over time.

Chromosome Changes

Another area of scientific study is a tumor's ability to lose or gain pieces of chromosomes. Each normal human cell has 23 pairs of chromosomes: 22 numbered pairs (1 through 22) and one pair of sex chromosomes. The most common chromosomal changes in brain tumors occur on chromosomes 1, 10, 12, 13, 17, 19 and 22.

- > Changes on **chromosomes 1 and 19** are most frequently found in **oligodendrogliomas**.
- > Changes on **chromosome 12** are frequently found in **glioblastoma**.
- > Changes on **chromosome 22** are most frequently found in **meningiomas**.

Scientists are studying how this information can best be used for diagnostic or treatment purposes.

The Cancer Genome Atlas (TCGA) Project

Studies of any specific gene are complicated due to the fact that there are many potential genes in the human genome to consider (the human genome has approximately 20,000 genes). While these genes interact with one another, they may also interact with environmental factors as well.

The Cancer Genome Atlas (TCGA) Project, funded by the National Cancer Institute (NCI) and the National Human Genome Research Institute (NHGRI), had a goal of completely cataloging all of the somatic genetic changes in more than 20 different cancers and making the data publicly available in order to improve the ability to diagnose, treat, and prevent cancer. TCGA started as a pilot project in 2006, prioritizing glioblastoma, ovarian cancer and lung cancer. This project was later expanded to include lower grade gliomas and several other cancer types.

The first GBM paper published under this project showed three critical biological pathways involved with glioblastoma. In a second glioblastoma paper published by TCGA, several factors associated with improved survival outcomes were identified. These include mutations in key genes involved in metabolism (IDH1 and IDH2) and high amounts of methylation (or “silencing”) of genes.

The first low-grade glioma paper published under this project found that two major genetic features can classify these tumors extremely accurately, including mutations of IDH1 or IDH2 genes, and “loss” of parts of chromosomes 1 and 19. These genetic features are now part of the standard process for diagnosing all gliomas, which are the most common type of malignant brain tumor in adults and are also quite common in children.

Since these publications, other scientists have described additional key genetic changes associated with malignant brain tumors. Several studies of medulloblastoma have identified that four distinct subtypes of these tumors are associated with specific age groups and specific clinical outcomes.

These types of large-scale studies have yet to be performed for non-malignant brain tumors.

Questions about Heredity

Concerns about heredity and brain tumors are common. If you have questions about your family history, we suggest the following:

- > Begin by sharing your family’s medical history with your primary physician. He or she will want to know the type of brain tumor and your relations to the person with the tumor. Although routine screening for brain tumors is not available as it is for breast or cervical cancer, unusual symptoms – such as headaches or short-term memory loss – can be investigated with your family history in mind.
- > If you have multiple family members diagnosed with brain tumors or have concerns about starting a family, consider a consultation with a genetic counselor. He or she can access the latest genetic information related to the specific tumor type in your family and advise you accordingly.
- > Share your family’s medical history with your kids and help them become good medical historians. Your children can learn their family history of brain tumors at the same time they learn about other diseases that run in the family, like high blood pressure or diabetes.

Chapter 5: Symptoms and Side Effects

The symptoms of a brain tumor are different in each person. While it is not possible to know exactly what symptoms to expect, understanding what might occur – and why – may help you better prepare for these possibilities. Symptoms like memory problems, seizures, or changes in personality or speech may be indications of a brain tumor. In some cases, tumors are discovered by accident, such as when a scan is performed for a non-brain tumor purpose, such as a head injury. It is possible that the tumor may not cause any symptoms that interfere with the normal workings of the body or the symptoms were so minimal that you were not aware of the tumor until a scan was done.

TUMOR EFFECTS

With a limited amount of space in the skull, the growth of anything that doesn't belong there can change the way the brain works. These changes may be temporary or permanent.

Tumors can cause direct damage to brain tissue, a shifting of the brain as the tumor grows or pressure on the brain. As a tumor grows, the symptoms often correspond to the affected parts of the brain. Therefore, it's important to ask your doctor where the tumor is located.

The most common tumor locations and the related effects are as follows:

- > **Frontal Tumors** cause a lack of interest in an individual's surroundings, as well as mood swings and changes in ethical standards. Problem solving may become difficult because of a lack of concentration. Behavior and personality changes also may occur, and short-term memory (memory of recent events) may diminish. When the memory cannot "remember" words, it may be difficult to express thoughts in words or writing. The frontal lobe also plans and begins the sequences of movement.
- > **Parietal Tumors** can cause a decreased awareness of sensation against the body. Difficulty recognizing body position or body parts may be noticed. If the tumor is in the dominant (usually left) hemisphere, confusion of the left and right sides of the body may occur. The parietal lobe also controls language and arithmetic ability. Numbers can be read, but the loss of recognition of left-right or up-down positioning can make it difficult to add, multiply or comprehend material presented in side-by-side columns. Similarly, sentences that contain a comparison or a cross-reference may not be understood.
- > **Temporal Tumors** are frequently "silent" unless they reach a significant size. They can cause a dreamy "déjà vu" state. Sense of time may be disturbed. The temporal lobe also controls the ability to hear and understand what is heard. Sounds or the source of the sound may not be recognized. Music and voices may be hallucinated or sounds may be heard louder or softer than they actually are. Behavior changes may occur. There also may be difficulty in remembering recent events.
- > **Occipital Tumors** disturb vision and the ability to recognize what is seen. The occipital lobe contains complicated visual connections; thus, a tumor in this area can cause various forms of visual loss. Loss of half of the vision in one or both eyes or blindness in only one direction may occur. Visual hallucinations may cause a temporary "dreamy" state. Facial expressions may not seem familiar.
- > **Optic Nerve Tumors** can reduce visual accuracy. The location of the tumor along the nerve determines what part of the visual field is lost. A tumor of the optic chiasm (where the optic nerves cross) can cause visual loss in both eyes. Headaches and nausea may be caused by pressure on the surrounding brain tissue.
- > **Cerebellar-Pontine Angle Tumors** (such as acoustic neuromas) cause pressure on the seventh and eighth cranial nerves. Ringing in the ears or a one-sided hearing loss (often first noticed when using the telephone) may occur. Dizziness and one-sided facial weakness are common. Vertigo, the sensation of the room spinning, is a common symptom from tumors in this area.
- > **Brain Stem Tumors** often cause vomiting and a clumsy gait (walking). The tumor can affect tongue movements, making swallowing and speaking difficult. One-sided hearing loss may occur. Unusual eye movements can cause dizziness or unsteadiness in walking. Double vision is common due to involvement of the third, fourth or sixth cranial nerves. Weakness of arms or legs may occur. Difficulty swallowing can occur with involvement of the lower brainstem.

- > **Hypothalamic and Pituitary Tumors** can disturb appetite and the desire for food. The pituitary gland also controls the normal production of hormones in the body. A tumor in this area may change the amount of hormones made by the pituitary gland. Hormone disturbances can cause water balance problems, abnormal growth, infertility, disruption of the menstrual cycle, hypertension, obesity, sleep disturbances and emotional changes. Sexual development may be delayed or advanced, or sexual desire may change.
- > **Thalamic Tumors** may cause sensory changes on one side of the body. Tremors during purposeful movement may be noticed.
- > **Posterior Fossa Tumors** (such as choroid plexus, fourth ventricle and cerebellar tumors) may cause tremors or a clumsy, uncoordinated pattern when walking. Nerve irritation may cause pain at the base of the head.

EMOTIONAL EFFECTS

When a brain tumor is diagnosed, it can take away your sense of security and control. This can be both unsettling and frightening. Uncertainty is among the most challenging things that you will have to grapple with on a day-to-day basis. The feeling that your body has betrayed you often leads to a rollercoaster of potent emotions.

Patients with a brain tumor often develop symptoms due to the stress of diagnosis and treatment. Decreased appetite, depression, irritability, fatigue, sleeplessness, temporary memory problems and restlessness are common. Nausea (a sick-to-your-stomach feeling), bladder problems or constipation also may occur. Your doctor usually can help you deal with these problems.

There is no “right” way for you to behave or feel when you have been given a brain tumor diagnosis. Dealing with changes to your appearance – such as shedding your hair or losing weight – and losing your one-time sense of invulnerability is difficult for the best of us.

It’s important to recognize the emotional effects of a brain tumor and find ways to cope. It may help to know that it is completely normal to feel a range of emotions when your life suddenly changes. Some people find that just having a loved one to talk to when days are difficult is enough. Others need some extra professional help, perhaps a caring member of your healthcare team, a social worker or a clinical psychologist. Support groups and relaxation exercises may also be useful. For information on coping strategies and tips on managing stress, see Chapter 9, “Coping.”

PHYSICAL EFFECTS

Treating a brain tumor takes a physical toll on one’s body. As you go through your treatment, know the potential side effects, such as hair loss or changes in your body. While the effects are different for every person, a brain tumor and subsequent treatment can change a person’s appearance, as well as their ability to carry on a full, active day.

Many hospitals offer make-up and hair sessions for those who have received cancer treatment. These seminars provide personal appearance tips and can help boost self-confidence. Oftentimes, when you think you look your best, you feel better about yourself.

People with a brain tumor often have questions about sex: “Can I still have sex? How soon after surgery can I have sex? Will my treatments affect my desire for sex?” Talk to a member of your healthcare team – they can answer your questions and provide suggestions. Your desire for sex may decrease temporarily because you may feel fatigued, unattractive or you may fear hurting yourself. Or, your partner may be overly cautious and afraid of hurting you. For the time being, consider replacing sexual activity with non-sexual physical closeness such as holding hands,

cuddling, kissing or hugging. Find activities you can comfortably share and special times to be alone.

Be realistic during your treatment – keeping up with your usual responsibilities may be too difficult. Medications, treatment and traveling to and from treatment can all cause fatigue. Set priorities. Do only what has to be done and if you still have the energy or inclination, then consider other chores or errands. Call upon friends and neighbors to help. Plan frequent rest periods during the day. Save your energy for special events or unavoidable chores.

It's also important for both patient and caregiver to continue treatment for any other medical conditions during this time and keep up with dental visits and eye care examinations.

More information on brain tumor types, treatments, clinical trials and other brain tumor resources can be found at www.abta.org or by calling 800-886-ABTA (2282).

Chapter 6: Diagnosis

Sometimes a brain tumor is found by accident – it may be seen on a scan performed for a non-brain tumor purpose – but most commonly, a tumor makes its presence known by interfering with the normal workings of the body.

Follow-up care for a brain tumor extends over a lifetime, not unlike many other medical conditions. At some point, depending on the type of tumor, your brain tumor may become a “chronic illness,” just as heart disease or diabetes are “chronic” conditions.

Understanding your tests – what they are, how they work and what they can or cannot show – can help you feel more comfortable and in control. If at any time you have questions about the tests ordered for you, feel free to ask. Your nurses and the professionals giving these tests can provide answers, fact sheets, helpful instructions and the reassurance you need to feel comfortable.

MAKING A DIAGNOSIS

Your doctor begins the diagnosis by taking your medical history and asking you to describe your symptoms, including how long you have had them, when they occur, the order of their appearance, if they seem to be brought on by something in particular and if they seem to be getting worse. Then your doctor will perform a basic neurological examination in the office.

NEUROLOGICAL EXAM

A basic neurological examination includes the following tests:

- > **Eye movement** by following a moving finger; pupil reaction and eye reflex using a pen light
- > **Vision**, including an examination of the optic nerve
- > **Hearing** using a ticking watch or tuning fork
- > **Reflex** using a rubber hammer
- > **Balance and coordination** by observing heel-to-toe walking, heel-to-shin movements, balance with feet together and eyes closed, rapid alternating movements such as touching the finger to the nose with eyes closed
- > **Sense of touch** using a sharp object and a cotton ball or paint brush
- > **Sense of smell** with various odors
- > **Facial muscle** including smiling and grimacing
- > **Tongue movement** and **gag reflex**
- > **Head movement**
- > **Mental status**, such as stating the current time and date, naming the current president
- > **Abstract thinking**, such as defining the meaning of “a stitch in time saves nine”
- > **Memory tests**, such as repeating a list of objects, describing the food you ate at yesterday’s breakfast, what occurred last month

If the results of your neurological examination lead the doctor to suspect you have a brain tumor, a scan will be ordered or you might be referred to a neurological specialist for additional testing, including scans, X-rays or laboratory tests.

IMAGING

Imaging takes the place of conventional X-rays, which do not show tumors located behind the hard bones of the skull or spine. The most commonly used imaging methods for diagnosis and follow-up are Computerized Tomography (CT) and Magnetic Resonance Imaging (MRI).

Both CTs and MRIs use computer graphics to create an image of the brain. During a scan, an injection of a special contrast material (dye) is given to make abnormal tissue more obvious. Contrast materials are able to highlight abnormalities such as tumors because the dye concentrates in diseased tissues due to the leakiness of blood vessels in and around brain tumors.

CT Scan

This scan combines an X-ray device with a computer. For some types of tumors, CT images are obtained both with and without contrast enhancement to provide important additional information.

If contrast is used, it is usually injected after a few pictures are taken. The patient lies on a table that slides into a doughnut-shaped opening. The CT scanner circles the head so the X-rays penetrate the brain from many directions. Absorption of the X-rays varies with the type of tissue being scanned. Thousands of thin cross-section readings are fed into the computer, which transforms the information into a picture. The CT scan is useful as a screening test in the emergency room; it can detect calcification, hemorrhage (bleeding), hydrocephalus (build up of fluid), edema (swelling) or shift. Recent studies on the radiation exposure caused during CT scans are helping doctors to redefine when a CT scan versus another type of scan is most appropriate.

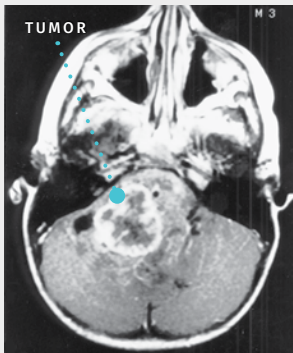
MRI Scan

The gold standard for brain tumor diagnosis, generally, is the MRI scan because it is more sensitive and specific. The MRI is a tunnel-shaped piece of equipment. Some pictures are taken before the contrast injection. If contrast is used, it will be injected prior to the completion of the scan. The patient lies on a table that slides into the tunnel. Inside the scanner, a magnetic field surrounds the head and a radio frequency pulse is introduced to the area. No X-rays are used. The magnetic field causes atoms in the brain to change direction. The radio frequency pulse causes another change of direction. When the pulse stops, the atoms relax and return to their original position. During relaxation, the atoms give off energy in differing amounts and at different intervals of time. Antennas pick up these signals and feed them into a computer, which assembles a picture. Because different atoms have their own characteristic radio signals, the computer can distinguish between healthy and diseased tissue.

Patients with certain cardiac monitors, pacemakers or some types of surgical clips cannot generally undergo MRI scanning because of the magnetic fields. For those who are claustrophobic, sedation or open MRI scanners may be an option.

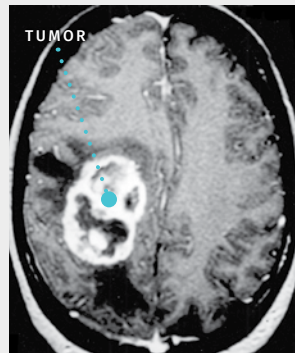
There are several different types of MRIs now available. Some of these are commonly used (such as fast MRI) while others are still being developed (such as diffusion tensor imaging).

MRI SCAN



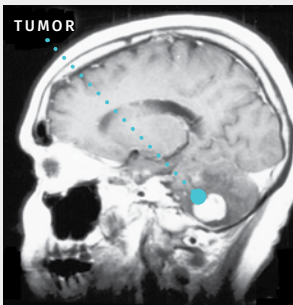
This MRI scan shows an ependymoma.

Scan courtesy of Dr. Regina Jakacki



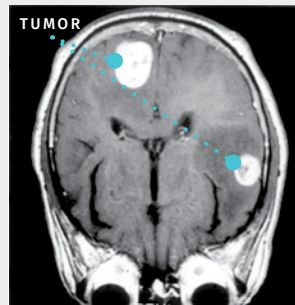
MRI scan of a glioblastoma multiforme.

Scan courtesy of Dr. Jeffrey Bruce



MRI scan of a single breast metastasis in the cerebellum.

Scan courtesy of Dr. Deborah Heros



MRI scan of multiple brain metastases.

Scan courtesy of Dr. Raymond Sawaya

An MRI offers images with excellent anatomical detail that provides clarity of the small structures in the brain, but the images often lack quantitative (or finely measurable) information. It is often difficult to determine the response to therapy because the side effects of treatment, especially radiation or immunotherapy, can cause inflammation or tissue death which can be difficult to distinguish from tumor progression. Researchers are working toward new scanning techniques that will more rapidly image treatment effects.

Cerebral Blood Volume (CBV) and Cerebral Blood Flow (CBF)

Some new scans measure the rate of blood flow into and through the brain. A contrast dye is given to the patient by intravenous (IV) infusion. The scanner begins taking pictures as soon as the dye is given. Using computerized timing, a succession of rapid pictures can be imaged, tracing the path of blood flow into the brain and to the brain tumor. These scans are currently used to help visualize the tumor's blood supply. New research indicates they may also be helpful as tools to monitor the effectiveness of treatments that affect tumor blood supply. These techniques are also used to scan spinal cord tumors.

These new methods are collectively called hemodynamic imaging. The information gathered can be converted into images or graphed into charts. Several different types of scanning equipment are used to produce these images: CT, MRI, PET and SPECT.

Dynamic CT and Dynamic MRI

The CT or MRI is combined with the ability to measure the uptake of the contrast dye from the time it begins to flow from the IV. Dynamic scans are especially useful in showing the growth of new blood vessels around a tumor.

fMRI (also called Fast MRI, Echoplanar, Real Time or functional MRI)

This technique produces MRI images in a faster sequence than traditional MRIs. The increased speed permits the tumor's use of oxygen to be depicted. Functional MRI may be useful prior to or during surgery to show the specific areas of the brain that control speech, movement and memory so they can be avoided.

Flow Sensitive MRI (FS MRI)

This type of scan combines functional MRI with images of cerebrospinal fluid (CSF) flow. FS MRI can be used to show the flow of CSF through the ventricles and spinal cord. It can be useful in planning for the surgical removal of a skull base tumor, spinal cord tumor or a tumor causing hydrocephalus (buildup of fluid).

Angiography and MRI Angiography (MRA).

Angiography is used to outline the presence and position of blood vessels in the brain. After injection of a contrast material into a deep artery, X-rays follow its flow through the blood vessels of the brain. MRI angiography, which is less invasive, uses a rapid succession of MRI scans to follow the blood flow and can be done with or without the injection of contrast dye.

The role of angiography for brain tumors is usually limited to planning the surgical removal of a tumor suspected of having a large blood supply or tumors growing into an area of the brain with an abundance of blood vessels. At times, angiography can be used as a means of embolizing or closing off large blood vessels that feed the tumor, making surgery easier.

Magnetic Resonance Spectroscopy (MRS)

Magnetic Resonance Spectroscopy produces images depicting function rather than shape. The equipment requires a special, highly complex facility.

Capable of measuring some byproducts of living tissue (called *metabolites*), this non-invasive scanning technique can depict patterns of activity that may be useful in diagnosing specific

tumors. MRS may be useful with low grade gliomas, tumors with a large amount of surrounding edema (swelling), and in differentiating between tumor recurrence and radiation necrosis (dead cells). This technique may also be valuable in suggesting the degree of malignancy.

Positron Emission Tomography (PET)

Positron Emission Tomography scans are not yet routinely used for diagnosis, but they can complement CT or MRI information by suggesting tumor grade. They are also used to distinguish between tumor regrowth, cells killed by radiation (necrosis) and scar tissue. Unlike CT or MRI scans, PET scans are quantitative (measurable). However, PET scans do not provide detailed images of the brain anatomy. To add anatomic detail, the latest PET scanners are being combined with CT or MRI scanners. In these hybrid scanners, PET and CT scans are acquired concurrently and the resulting PET image is fused with the CT image. The use of PET in brain tumor studies is increasing as scientists develop new imaging drugs, smaller and more mobile PET facilities, and as PET scanning is combined with other types of scans.

In a PET scan, a low-dose of a radioactive substance is injected into the patient. The PET scanner has a circular detector into which the patient's head or body is moved to detect the amount of the radioactive substance taken up by various parts of the brain. The most commonly used radioactive substance for tumor imaging is a radioactive sugar (FDG). The FDG has been most commonly used because a growing tumor consumes sugar at a high rate; radiation necrosis or scar tissue consumes almost no sugar. However, the normal brain itself consumes a lot of sugar causing considerable background color in the PET images. Other radioactive substances that are now in the early phases of clinical development may provide a clearer picture of the tumor as well as the ability to capture additional details about the tumor or the activity of the tumor cells.

In PET scans, measurements of brain or brain tumor activity, determined by concentrations of the radioactive substance, are fed into a computer that produces a color-coded moving picture of the brain as it accumulates the FDG.

In the past, the use of PET had been somewhat limited because the equipment is expensive and requires radioactive materials (drugs) to be synthesized on-site. As new radioactive substances become available, an increasing number of facilities now offer or can arrange PET scanning. Truck-mounted mobile PET and combination PET/CT scanners are also bringing this technology into more hospitals.

Single Photon Emission Computerized Tomography (SPECT)

Single Photon Emission Computerized Tomography is not routinely used in the initial diagnosis of brain tumors, but might complement information obtained from other scans.

A SPECT scan is similar to PET because it monitors how the brain reacts to and uses radioactive-tagged materials. In a SPECT scan, a special camera measures the rate of emission of the material as it moves through the brain, and then images are generated from that information. After MRI or CT scans, this test might be helpful in distinguishing between low-grade and high-grade tumors or between recurrent tumors and necrosis (dead tumor cells).

Magnetoencephalography (MEG)

A MEG scan measures the magnetic fields created by nerve cells as they produce the small electrical currents used for neurotransmission. No physical contact is required to record the signals. The images created during the scan help scientists identify how parts of the brain interact with each other, how the brain processes information, and how the pathways function as information enters the brain. This may also help us understand why certain brain tumors, based on their location, cause specific functional problems.

The device used during a MEG scan looks like an old-fashioned hair dryer. When the patient moves, a computer-generated image shows which brain area is responsible for directing the motion.

The MEG images are used in combination with information from other types of scans to determine the function of specific areas of the brain. MEG scanning is available at a limited number of facilities.

X-RAYS

Plain skull X-rays are usually not necessary for diagnosis except to help determine if calcification or bone erosion is present. (Slow growing tumors can cause calcification and increased intracranial pressure might cause erosion.) An X-ray image might be used to determine the condition of the skull next to meningeal and skull base tumors.

The science of imaging brain tumors is a rapidly changing field. Increasing image resolution, new contrast dyes, the ability to attach scannable tracers to drugs and methods of rapidly determining treatment effectiveness all hold promise for the future of brain tumor therapy.

A radiologist interprets the computer images produced by scans and X-rays. The pictures help establish a tentative diagnosis and might suggest the type of tumor, but they are not definitive. An exact diagnosis can only be made by examining a sample of the tumor tissue under a microscope. The pathology report is often enriched with next-generation analysis of genomic alterations.

LABORATORY TESTS

Biomarker Research

Recent advances in the scientific ability to detect proteins or DNA shed by brain tumor cells in bodily fluids has given rise to an area of science called biomarker research. These miniscule bits of material are being explored for their potential use in diagnosis and treatment, as well as monitoring the effectiveness of treatments. To date, biomarkers have been identified in blood, plasma, cerebrospinal fluid, urine and saliva.

While the science of these findings is advancing rapidly, their practical, everyday use in a clinical setting is still very unclear and requires large clinical trials. Biomarker tests that predict the likelihood of survival over a period of time and tests that indicate the aggressiveness of tumor cells are now making their way into hospitals. Biomarker research is forming the basis for individual and personalized medicine. This new and fascinating area of study is in its infancy across all fields of medicine.

Lumbar Puncture (Spinal Tap)

Lumbar puncture is used to obtain a sample of cerebrospinal fluid (CSF). This procedure is usually avoided if there is any indication of increased intracranial pressure because of the risk of *herniation*, or the brain's bulging through an opening in a membrane, muscle or bone.

The sample of CSF is examined in a laboratory to determine if tumor cells, infection, protein or blood is present. This information is particularly helpful in diagnosing primary CNS lymphoma, a pineal region or metastatic tumor. The presence of tumor cells in the CSF indicates tumor spread. That information is used for tumor staging and helps the doctor determine appropriate treatment choices.

In addition to tumor cells and substances that indicate the presence of a tumor, the CSF may also be examined for the presence of known tumor markers. Some common biomarkers include the following:

- > MGMT
- > EGFR
- > IDH1/IDH2
- > EGFRvIII
- > 1p/19q
- > TERT

Researchers continue to explore and validate biomarkers for different tumor types.

Myelogram

A lumbar puncture is used to inject a special dye before a myelogram. The patient is then tilted to allow the dye to mix with the spinal fluid. This test is used primarily to diagnose a spinal tumor and obtain pre-operative information for spinal tumor surgery.

Spinal MRI has replaced myelography for most conditions.

Evoked-Potentials

Evoked-potential testing uses small electrodes to measure the electrical activity of a nerve. This test is particularly useful in detecting a vestibular schwannoma (acoustic neuroma).

Evoked-potentials can also be used to monitor neurological function during the surgical removal of a tumor.

Audiometry

This hearing test is useful in the diagnosis of a cerebellopontine angle tumor, such as the vestibular schwannoma (acoustic neuroma).

Endocrine Evaluation

Measurements of hormone levels in samples of blood and urine are used, along with scans, to diagnose a pituitary or hypothalamic tumor.

Perimetry

This technique measures the size of visual fields. The information obtained might be useful in diagnosing a tumor in the area of the optic chiasm, such as a pituitary tumor, or anywhere along the optic pathways, such as the optic radiations for a parietal, temporal, or occipital tumor.

BIOPSY

A biopsy is a surgical procedure in which a small amount of tumor tissue is removed. The neurosurgeon submits the tumor tissue to a pathologist for analysis. Only then is a tumor diagnosis possible.

A biopsy can be performed as part of the surgery to remove the tumor or as a separate diagnostic procedure.

For areas considered inoperable, the surgeon is often able to perform a *needle biopsy* through a small hole drilled into the skull, called a *burr hole*. A narrow, hollow needle is inserted through the burr hole and tumor tissue is removed from the core of the needle.

Stereotactic biopsy is a computer directed needle biopsy. The computer, using information from a CT or MRI scan, provides precise information about a tumor's location and its position relative to the many structures in the brain. Stereotactically-guided equipment might be moved into the burr hole to remove a sample of the tumor. This is called a *closed biopsy*.

When a biopsy is not performed, diagnosis relies solely on the interpretation of other test results.

TUMOR STAGING

Staging determines if a tumor has spread beyond the site of its origin. In cancers such as breast, colon or prostate this is primarily accomplished by a pathologist’s examination of nearby tissue, such as lymph nodes. In those cancers, staging is a basic part of the diagnostic work-up.

Staging for central nervous system (CNS) tumors is usually inferred from CT scan or MRI images or by examining the cerebrospinal fluid. Scans taken after surgery are used to determine if there is remaining tumor. CNS tumors that are especially prone to spread are studied with both scan images and laboratory tests. For example, patients with medulloblastoma will often have their cerebrospinal fluid examined for the presence of tumor cells. Those patients will also have scans of their spinal cord because of that tumor’s tendency to spread to that location.

Staging information often influences treatment recommendations and prognosis.

CHANGE OF DIAGNOSIS

Although it may initially seem alarming, your diagnosis and the name of your tumor might change. There are several factors that might cause the change in diagnosis:

- > Tumors do not always remain static. They can undergo transformation, usually to a higher grade. If that occurs, the name and grade of the tumor might change. A grade III anaplastic/malignant astrocytoma could become a glioblastoma (also called a grade IV astrocytoma).
- > Inspecting only a small sample of the tumor, such as that obtained by a needle biopsy, might not be representative of the whole tumor.
- > As scientists learn more about the biology of brain tumors, they are becoming aware of the molecular differences and similarities in tumors. Sometimes this means renaming or regrouping tumors.

HORMONE	RESPONSIBLE FOR	NORMAL BLOOD LEVEL IN ADULTS
ACTH adrenocorticotrophic hormone	Production of cortisol – a natural steroid needed to control blood pressure, sugar and salt levels	9 to 52 pg/ml
GH growth hormone	Controls bone growth; height; body proportion in the extremities and jaw	0 to 3 ng/ml
PRL prolactin	Controls milk production in women, impacts sex drive and sperm counts in men	Males and non-pregnant women: 0 to 20 ng/ml In pregnancy: 10 to 300 ng/ml
TSH thyroid stimulating hormone	Controls thyroid functions such as metabolism, heart rate and appetite	0.2 to 4.7 mcU/ml

All grading systems have inherent difficulties and are not precise.

- > Criteria used to assign grades can be subject to interpretation by each pathologist.
- > Tumors are not uniform and the sample examined might not be representative of the entire tumor.

ABOUT LESIONS

Lesion is a general term which refers to any change (abnormality) in tissue. Tumor, inflammation, blood, infection, scar tissue or necrosis (dead cells) are all examples of lesions that may be found in the brain. Determining the nature of the lesion is the work of the pathologist.

If your doctor tells you a lesion was seen on your scan, the next step is to ask your doctor what type of lesion she or he believes this to be. Treatment will be determined based on the type of lesion.

ABOUT FOLLOW-UP TESTING

At intervals during and after treatment, your doctor will probably order some of the same tests you took when your tumor was first diagnosed. These tests will be used to see if the tumor has disappeared, is shrinking, remains the same or has changed.

For many patients, a first follow-up MRI scan will be done one to three months after surgery or after the completion of radiation therapy. This gives the brain a chance to begin healing from the effects of surgery or radiation. Although it can be difficult to wait, scans done immediately after treatment would most likely show the swelling that can occur as a result of surgery and radiation and would not be truly representative of the status of the tumor itself.

Following that initial post-treatment scan, your doctor will determine how often you should have follow-up scans. Depending on the type of tumor, your doctor may suggest MRIs every two or three months, six months or perhaps yearly. The follow-up may continue even 10 or more years after treatment. Very late recurrences can happen. A doctor who knows your history can determine if new symptoms are related to the tumor or to another medical condition.

Scans help to measure the effectiveness of the treatment and monitor for possible recurrence.

Your doctor will tell you when your next scans or tests should be done. If you do not have this information, call your doctor's office and ask. Your follow-up is as important as your treatment.

PROGNOSIS

A prognosis is an educated guess about the future course of a disease in a specific individual. It is not the same as a prediction, which accurately forecasts the future. Prognosis is based on multiple factors.

For example, prognosis is based on the type of tumor, its grade, location and spread (if any); the age of the patient; how long the patient had symptoms before the tumor was diagnosed; how much the tumor has affected the patient's ability to function; and the extent of surgery, if surgery was performed.

The availability of successful therapies also influences the prognosis. A non-malignant tumor located in a vital area may be life threatening, while certain malignant tumors may be successfully treated with radiation or chemotherapy.

WHAT HAPPENS NEXT

Your mind is probably racing with thoughts and filled with questions. One way to help stay organized and in control is to write your questions in a notebook. Try listing your questions by placing important questions near the top or by numbering the questions in order of importance. This list will help ensure that your concerns are addressed by helping you stay organized and focused.

It can also be helpful to have a friend or family member accompany you to your appointment. Not only can they offer comfort and moral support, they can help make sure that your questions are being asked and answered by checking them off from the list and writing down responses.

If it is OK with your doctor, bring a recording device with you. Then, you can listen to the doctor again in the comfort of your own home.

During your visits to the doctor, ask for written information about your brain tumor, your symptoms, suggested treatments and your medications.

Some insurance companies require pre-certification of scans. Check your policy for guidelines. If you call for pre-certification, record the date, the name of the representative to whom you speak and their response.

Before you leave the doctor's office, make sure you understand any instructions that were given. For example, do you have another appointment? If so, when is it? If you are scheduled to have additional tests, do you know why, when and where to go? Ask the doctor or nurse to write important dates and instructions in your notebook.

QUESTIONS TO ASK

Whether you are just beginning treatment, are a long term survivor, or are somewhere in between, you probably have some unasked or unanswered questions. You might be concerned about your symptoms or want to ask about treatment options. You may have obtained copies of your medical records and read something you do not understand. Or perhaps you would like guidance about resuming your routine activities.

We encourage you to take these questions to your healthcare team. Your doctors and nurses can respond with personalized answers which cannot – and often should not – be provided by outside sources. By asking questions, you are participating in your health care. By gathering information, you will feel more comfortable making decisions about your treatment plan.

In this section, we offer some sample questions you may want to ask at various times during your brain tumor journey. Feel free to modify this list based on your particular concerns and situation.

Following diagnosis, most questions focus on the disease and its treatment. Later, they often include concerns about daily activities, nutrition, rehabilitation or medications. If a topic causes concern at any point during care, it deserves to be addressed.

Questions to ask your healthcare team may include:

- > Where is the tumor located?
- > Based on the scans, do you have an idea of the type of tumor?
- > What do we do next? Do I need more tests? Do I need to see any specialists?
- > Until we know more, can daily routines continue? Driving a car? Exercising? Working?
- > Do I need to take any medication? If so, what is it for? What are the side effects?
- > Can the tumor be operated on?
- > What are the risks in removing this tumor?
- > Are there treatments other than surgery?
- > If the tumor cannot be operated on, what treatment is recommended?
- > Can treatment wait? How long?
- > What might happen without treatment?

Keep your notebook handy to record answers to your questions. Asking and answering questions is essential to participating in your own care.

If treatment requires a hospital stay, ask the doctor for the name of the hospital. Many doctors are on staff at more than one facility, so there may be options. Once the treatment and location are finalized, the name of the hospital, the hospital department and the day and time of the appointment should be recorded in the notebook. Ask the doctor if any papers should be brought to the hospital. Also, check with your insurance company; they may need to pre-certify your treatment or hospitalization.

SECOND OPINIONS AND SEEING A SPECIALIST

After diagnosis, one of your next visits will likely be to a specialist. Regardless of whether the next step is a consultation regarding surgery, radiation, chemotherapy or another treatment, the basic questions are very much the same.

For many patients and their families, speaking to another doctor may help to better understand the disease and treatment options. In some instances, an insurance company may require a second opinion before they will cover a surgical procedure. Either way, second opinions are considered standard medical practice today. However, before seeking a second opinion, make sure it is safe to delay treatment.

There are several ways to obtain a second opinion. Many doctors are willing to suggest a specialist and may even make the appointment. Most hospitals offer physician referral services that can identify medical staff with expertise in treating brain tumors. Many patients receive referrals from family and friends. Additionally, the ABTA has a listing of well-regarded brain tumor treatment centers throughout the country. Visit www.abta.org, call 800-886-ABTA, or email abtacares@abta.org.

Some insurance policies require patients to stay within a network of providers. If so, ask your insurer for a list of in-network doctors who specialize in the treatment of brain tumors. An out-of-network doctor may result in additional costs and/or other responsibilities for the policy holder. It is important to know what those obligations would be before selecting a doctor.

Once you select the consulting doctor(s), call their office and ask about their procedure for obtaining a second opinion. Some doctors will review medical records and scans without the patient being present. Others will ask to see the patient too. The doctor's office can provide a list of the medical records needed to render an opinion and the best way to have them delivered.

Questions for a Specialist

You will want to know:

- > What treatment is recommended?
- > What is the goal of that treatment? To cure the tumor, to control the tumor or to control symptoms?
- > What are the potential benefits of the treatment?
- > What are the risks and side effects of the treatment?
- > What will happen if I don't have this treatment or if I postpone it?
- > Are there other options beside this treatment?
- > Is this an experimental treatment?
- > Will I need any more tests before the treatment begins?
- > How will we know if the treatment was effective?
- > What type of follow-up will I need and when?

OBTAINING MEDICAL RECORDS

Medical records are needed for a second opinion. However, the laws for obtaining records vary by state. Many states allow medical records to be released directly to the patient. Other states require that medical records be sent directly to the consulting physician.

The quickest way to obtain these records is from the doctor's office. Copies of scans, pathology, operative, consultation reports and office visit records are generally available from the doctor's office. Your doctor may be able to access your scans electronically and copy them for you. There may be a charge for providing a copy of medical records.

The consulting doctor will also ask to see the actual scans (not just the reports). If your doctor does not have the scans, call the hospital radiology or imaging department where the scan was done. MRI, CT, fMRI and PET scans can all be copied. **Never send an original through the mail.** Since there is a sizeable fee to copy scans, get a list of the exact scans needed.

Some hospitals keep reports and scans in electronic or digital files. These files can be copied onto a CD or DVD, transferred electronically or printed as traditional records. The consulting doctor can tell you which version is preferred.

Some consulting doctors (and some patients) ask for a second reading of the pathology slides. To obtain your slides, call the pathology department of the hospital where the surgery was performed.

Some slides can be copied. If they cannot be copied, ask if the hospital has a "paraffin block" sample of your tumor tissue. This is a larger piece of tissue, stored in a wax base, from which new slides may be created. Paraffin blocks, due to their size, are usually kept only a few years. Slides may be kept longer. Again, there may be a charge to duplicate or ship the pathology slides.

Written records of hospitalizations are kept in the medical records department of the hospital for several years. Afterward, they may be copied into electronic files or otherwise archived.

Before providing copies of hospital records, the medical records department will ask for the patient's:

- > Name (the name under which the patient was hospitalized)
- > Birth date
- > Social security number
- > Approximate date of hospitalization
- > List of requested reports

Some hospitals require a written authorization from the patient to release the records.

Find out if the records will be released to the patient or sent directly to the consulting doctor. If the records need to be picked up, find out where and when they will be available. If the records are to be forwarded, the doctor or hospital will need the consulting doctor's name, address and telephone number. Find out when the records will be sent and by what method. It may be best to have the records shipped by a company that has a tracking system for packages.

Finally, the consulting doctor should be alerted as to when the records will arrive. The doctor should be asked if he or she will provide the consultation by phone or if an appointment is required. The doctor will need enough time to appropriately review the records before rendering an opinion.

Whether you are the patient or a family member, it is tempting to read through medical records before sending them on to a consulting doctor. However, these records are written in technical medical terms. The words and terms may be very new to you and may be alarming because they can be difficult to understand. Questions that arise about anything you find in the records can be written in your notebook and discussed with your healthcare team.

ABOUT INSURANCE

After your first visit, you will need to verify your healthcare insurance coverage. The answers to most of your insurance questions can be found in the insurance policy itself or the policy manual. If you do not have a copy, now is the time to obtain one.

For employer-provided health insurance, contact your employer's Human Resources department or your benefits manager and ask for the manual. For individual policies, call your insurance agent. For Medicare/Medicaid coverage, call the Medicare Hotline at 1-800-633-4227. For CHIP (Comprehensive Health Insurance Programs) coverage, contact your state department of insurance.

If you are uninsured, begin by contacting the social worker at the hospital at which you will be treated. You can reach the social work department by calling the general hospital number and asking for the social work office. The social worker can outline federal assistance programs, local and national funding organizations and ways to help you obtain alternate forms of health care coverage.

Questions for Your Insurance Provider

Be sure you know the answers to these questions:

- > Do you need to obtain pre-certification for hospitalization or treatment? If so, who do you call? Most insurers include the pre-certification telephone number on the back of the insurance card. When you call, be sure to record the name of the person you speak with, the date and the "case number" assigned to your claim.
- > Do you need to obtain a second opinion before non-emergency surgery? If so, are there any limitations on who provides the second opinion?
- > Do you need to stay within a particular network of hospitals or physicians to receive your benefits? Do you have a current list of those providers? What will happen if you are treated "outside the network?"
- > Does your policy have a deductible? If so, how much of that deductible have you paid for the year? Knowing this will help avoid "surprise" bills for which you are responsible.
- > Will your insurance cover investigational treatment if you choose it?

MORE INFORMATION ON BRAIN TUMOR DIAGNOSIS, IMAGING, INSURANCE, SECOND OPINIONS AND OTHER BRAIN TUMOR RESOURCES CAN BE FOUND AT [ABTA.ORG](https://www.abta.org)

Chapter 7: Seizures

Seizures are common symptoms of a brain tumor. Between 25 and 40 percent of people diagnosed with a brain tumor will have a seizure at some time during their illness. A seizure can be a startling experience; however, understanding what a seizure is and what to do if one should occur can minimize fear and potential injury.

For some people, a seizure may be the first clue that something unusual is happening in their brain. Seizures might be caused by a brain tumor or by the surgery to remove it. Seizures can also be totally unrelated to a brain tumor. For example, an injury to the head, a stroke, alcohol or drug withdrawal, or a fever can all cause seizures. Or, the cause can sometimes be unknown.

Most seizures can be controlled with medications called antiepileptic drugs (AEDs). Surgery or a ketogenic diet are also sometimes used to help treat ongoing seizures.

This chapter provides information and resources to help people affected by seizures understand what they are experiencing and to learn how to live with this symptom.

WHAT ARE SEIZURES?

A seizure is an episode of abnormal electrical activity in the brain.

During normal brain activity, the body's nerve cells communicate with each other through carefully controlled electric-like signals. Those nerve cells send thousands of signals back and forth, giving instructions to all parts of the body. If something interferes with those signals and they become more intense, a seizure results.

Some events may trigger seizures. Bright lights, flashing lights, specific odors, lack of sleep, missed meals, increased stress, alcohol, new medication or changed medication doses can all be triggers. To help identify your personal triggers, keep a diary or journal of activities and feelings that occur prior to each seizure.

What does someone having a seizure look like? For some, jerky shaking and trembling may be the first thing you see. Sometimes these body movements become extreme. In addition to unusual body movements, there may be a change in awareness, unusual sounds, visions or sensations. The type of seizure depends on which part of the brain is experiencing the abnormal electrical signals.

Other times, a seizure may be as simple as someone appearing to be "far away." For others, when they are having a seizure, they experience hearing music only they can hear or a phrase chanted repeatedly until the seizure is over.

The person having the seizure will not be aware of others around them during the seizure and may not respond to touch or hearing their name. These are all normal variations of seizures. Although seizures are usually brief, their effects may linger for several hours. Recurrent seizures are referred to as epilepsy.

Having a seizure does not automatically mean your tumor is growing. If you experience a seizure after a long period of being seizure free, share this information with your healthcare team. They can best advise you as to your next steps in identifying the significance of the new seizure activity.

SEIZURE WARNING SIGNS

Most seizures occur randomly and without any particular cause. However, you might have some advance notice. Learning these signals, called auras, can help you prepare for a seizure. A headache, mood change, muscle twitch, dizziness or a particular smell are auras that may happen a few seconds or minutes before the actual seizure and might signal a coming seizure. Use that time to safeguard yourself. If you are chewing, remove the food from your mouth. If you are walking, sit or lie down. If you are with someone experiencing an aura, assist them in finding a safe place.

How to Help Someone During a Seizure

Most people have never seen anyone have a seizure. It is normal to feel concerned or anxious about the possibility. Learning what to do, in advance, may help calm some of those fears. Sharing this information with your family or friends can help prepare them as well. Remember that most seizures end naturally.

If you are with someone who has a seizure, try to remain calm and make sure to protect the person from environmental harm. Most of the time, a person having a seizure requires no assistance other than a caring presence and observation.

First, make sure the person is breathing. Loosen clothes around the neck. Move pillows, blankets or any other items away from the nose and mouth. If the person is having trouble breathing,

immediately call for emergency help. Do not place anything in the person's mouth as this could obstruct their airway.

If the person appears to be breathing well on their own, clear the area of sharp objects or anything else that could be dangerous. Remove the person's eyeglasses. If possible, help the person lie on their side. This helps keep their airway open. Protect the patient's head from being bumped if they are having a generalized seizure. Do not attempt to restrain a person's arms or legs during a seizure as this may result in an injury. Do not put anything in the person's mouth, including your fingers. During a seizure, anything placed in the mouth will block the airway and cause breathing problems. Also, as the jaw often clenches during a seizure, your fingers could be bitten.

Most seizures last several minutes. After the seizure ends, allow time for the person to recover. They may be confused for a few moments. This is normal. Help re-orient them. Tell them who you are, where they are and what happened. Help them find a place to rest until they have recovered.

Call for emergency assistance if:

- > The person is having difficulty breathing
- > The person injures himself or herself
- > The seizure lasts more than 5 minutes
- > A second seizure immediately follows
- > The seizure occurs in water
- > The person is pregnant or has diabetes

TYPES OF SEIZURES

There are two primary types of seizures – *partial* and *generalized*. The type you experience depends on which area of the brain has the abnormal electrical signals.

Partial Seizures

There are two types of partial seizures – *simple* and *complex*.

> *Simple Partial Seizures*

Simple partial seizures commonly cause jerking or twitching (if the frontal lobe is involved), tingling or numbness (if the parietal lobe is involved) or other sensations. These symptoms can begin in one part of the body and then spread to other areas. Chewing movements or lip smacking (if the anterior temporal lobe is involved), buzzing in the ears, flashes of lights, sweating, flushing and pupil dilation are other common symptoms. Psychic symptoms include a sense of déjà vu, imaginary sights (if the occipital lobe is involved), smells (if the temporal lobe is involved), tastes or imaginary sounds. Simple partial seizures do not cause unconsciousness.

> *Complex Partial Seizures*

Complex partial seizures cause some loss of consciousness and usually indicate temporal lobe involvement. Uncontrolled body movements might occur. The seizure may be preceded by, accompanied by or followed by psychic symptoms. Psychic symptoms include a sense of déjà vu, imaginary sights (if the occipital lobe is involved), smells (if the temporal lobe is involved), tastes or imaginary sounds. A state of confusion may continue after the seizure activity. In patients with low-grade gliomas, this is the most common type of seizure.

Generalized Seizures

These seizures may begin as partial seizures and abruptly change into generalized seizures. There are several different types of generalized seizures.

> *Absence (Petit Mal) Seizures*

Absence seizures cause a brief delay in consciousness and may be accompanied by a feeling of limpness. The person having the seizure may miss a few words or stop speaking for a few seconds during a conversation. It may look like daydreaming. The beginning and end of the episode is usually sudden. This type of seizure most commonly begins in childhood and often stops by young adulthood.

> *Atypical Absence Seizures*

Atypical absence seizures may cause more extensive changes in muscle tone or they may have a more gradual beginning and ending than typical absence seizures.

> *Atonic Seizures (Drop Attacks)*

Atonic seizures are characterized by sudden limpness. Generally, all muscle tone and consciousness is lost.

> *Myoclonic Seizures*

Myoclonic seizures cause single or multiple muscle twitches, jerks or spasms.

> *Tonic-Clonic (Grand Mal) Seizures*

Tonic-clonic seizures are common in people with low-grade gliomas, but can occur with any type of glioma. The seizure involves a sudden outburst, then a loss of consciousness, followed by twitching and relaxing muscle contractions. The person might bite their tongue, lose control of body functions and take very shallow breaths. This usually lasts for two or three minutes and is followed by limpness. When the person regains consciousness, they may be sleepy, have a headache, be confused and/or have sore muscles. Most people are able to return to their normal activities after resting. If the seizure begins again, call for emergency assistance.

SEIZURE TREATMENT

Seizures may be controlled in three ways. Sometimes, a combination of methods is used.

Medications

Antiepileptic drugs (AEDs) are the most widely used method of controlling seizures. They are prescribed to prevent seizures or to decrease their frequency. There are different types of AEDs – the type your doctor prescribes for you depends on your seizure history and the type of seizures you experience.

Surgery

Surgery to remove the tumor may also stop or help control your seizures. Using sophisticated brain mapping techniques, a neurosurgeon may be able to define the exact area of the brain causing the seizures and surgically remove it.

Ketogenic Diet

The ketogenic diet is a high fat, low carbohydrate diet that may help control ongoing seizures that do not respond to seizure medications. A doctor should carefully prescribe the balance and components of your daily food intake and the diet must be carefully followed on a daily basis. Dietitians may also recommend necessary vitamin and mineral supplements. Blood tests and close monitoring are used to watch for side effects and to verify effectiveness of the diet.

The ketogenic diet is primarily used to treat children for whom seizure medications are not effective. Some children combine the diet with lower doses of seizure medications. Although adults could follow the diet, AEDs tend to be prescribed first due to the diet's very strict food restrictions.

USING ANTIPILEPTIC DRUGS

The goal of drug therapy is always to control seizures with the lowest effective doses of antiepileptic medication and with the least side effects. There are several important points to remember:

Maintain a Steady Level

Antiepileptic medications work best when there is a steady level of the drug in the body. The drug needs to reach and remain at the ideal level to be effective. Medication must also be taken regularly and as prescribed. If you miss a dose, do not take a double dosage. Instead, resume your regular schedule and notify your doctor. If you stop taking your medicine abruptly, seizure activity will increase. Call your doctor for assistance if you miss more than one dose, if you notice an increase in your seizures or if you develop a rash.

Check Levels if Indicated

Some medications require frequent blood tests in order to check the drug levels in the body. Ask your doctor if the medication should be monitored in this way and, if so, find out where and when to have those blood tests done. The medications might be adjusted based on the results.

Minimize Possible Drug Interactions

Many prescription and over-the-counter medications can influence the effectiveness of AEDs. Inform your doctor of all medications you are taking, including vitamins, nutritional supplements and herbal remedies.

Alcohol also interferes with some antiepileptic drugs. Check with your doctor before consuming alcohol.

Risks and Guidelines

Your doctor or nurse will tell you how long you will need to take the antiepileptic medications. The decision is based on seizure history, how often seizures occur, your MRI scans, EEG (electroencephalogram) results and your treatments. The decision to taper off antiepileptic medication should be carefully planned by the doctor and the patient, with all appropriate precautions taken. Medication should never be abruptly stopped without a doctor's approval, as it may trigger a seizure.

If one medication does not control your seizures, another drug or a combination of drugs may be prescribed.

TIPS FOR MANAGING COMMON AED SIDE EFFECTS

Discuss side effects with your doctor – especially if they persist and do not feel manageable. The following information may help you manage some common side effects of seizure medications.

Drowsiness or Dizziness

If you are experiencing drowsiness or dizziness as a result of your AEDs, do not operate equipment or machinery, and do not drink alcoholic beverages. Use caution on stairways. Install grab bars in the shower and next to the toilet (these can be rented from a medical supply store).

Gum Swelling

Some AEDs can cause your gums to become inflamed, red, swollen, tender or bleeding. This side effect is influenced by bacteria levels in the mouth, so it is important to maintain good oral

hygiene with regular brushing and flossing. If your gums are swollen, try using a soft tooth brush or a mouth care sponge (available at most drug stores). Avoid mouthwashes containing alcohol that may burn and irritate your gums. Baking soda-based mouth rinses may provide relief. Be sure to tell your dentist about your medication. Frequent professional cleanings may help limit gum swelling.

Rash

If you notice a rash after starting AEDs, notify your doctor immediately. A rash can indicate an allergic reaction to the seizure medication or may be due to an increased drug level. If itching accompanies the rash, a cool shower may provide relief by constricting the blood vessels in the outer layer of your skin. Pat your skin dry instead of rubbing. Do not use lotions on the rash unless your doctor or nurse suggests it. Do not take additional doses of the medication that may be causing the rash until you have spoken with your doctor.

Bone Disorder

Long-term use of seizure medications may cause bone disorders. The amount of calcium in the bone may decrease causing brittle bones and fractures. Decreased levels of vitamin D and phosphorus may also contribute to this side effect. Bone and blood tests can monitor these conditions and supplementation may be recommended. Regular exercise also supports healthy bones.

Nausea and Vomiting

Take your medication with meals to decrease stomach upset. If stomach upset continues, ask your doctor about anti-nausea (antiemetic) medication. Do not use over-the-counter antacids or aspirin-containing preparations for upset stomachs without first checking with your doctor since they may interfere with some seizure medications.

Continued Seizures

Some seizures simply do not respond to a given medication, and you may have to try another medication. Flu vaccines, prescription and non-prescription drugs can increase seizure activity. If you suspect that you are experiencing this problem, make a list of all your medications and share it with your doctor or pharmacist. Be sure to let your doctor know the frequency and type of your seizures and if the side effects of a particular drug interfere with your quality of life. Discuss this with your doctor and ask about other options for controlling your seizures.

Questions for Your Doctor or Nurse about Seizure Medications

- > What is the name of the seizure medication you have prescribed?
- > Why did you choose that particular medication for me?
- > How much do I need to take and how often?
- > Do I need to have any tests to monitor the medication in my blood and body?

Notify Your Doctor Immediately If You:

- > Have any difficulty breathing
- > Run a temperature
- > Notice the whites of your eyes appear yellow
- > Have tiny purple spots on your skin
- > Develop a rash
- > Become unusually confused
- > Have difficulty urinating
- > Bruise easily

Chest pain or inability to awaken someone taking seizure medication may be a medical emergency.

- > Is there anything that might interfere with its effectiveness, such as other medications or natural products?
- > What are the most common side effects?
- > What are less common side effects?
- > What side effects should I call the doctor about?
- > What side effects lessen with time?
- > What strategies do you recommend for managing side effects?
- > What precautions do I need to take due to the seizure medication?
- > Can I drive a car? If not, for how long?
- > Is there anything else I can do to minimize or control the seizures, enhance the effectiveness of seizure medication and/or lower the dose of the seizure medication?
- > What length of time do you anticipate that I will need to take seizure medication?
- > What tests do you use to evaluate if I need to continue taking the seizure medication over time?

LIVING WITH SEIZURES

Seizures are generally unpredictable. Here are a few suggestions for managing your seizure disorder.

Stress

Stress is a cause of seizures. You may be able to reduce stress through exercise, meditation, yoga, guided imagery, deep breathing or coping skills training. Discussing your seizure disorder with family and friends may help diminish some of the stress. Talking with others who have a seizure disorder or with a professional counselor can help you feel less isolated. Another option is to consult a neuropsychologist, a professional trained in the workings of the brain and the psychological impact that neurological disorders can have on a patient and their family.

Relationships

Seizures can be stressful for you, and they can also affect your relationships with family and friends. Communicating openly with your family and friends may help diminish some of the stress seizures can cause. You may feel afraid of having a seizure around other people or you may feel “different” because of your seizures. Talking with others who have seizures can help you feel less isolated. A professional counselor can help you with lifestyle adjustments.

Driving

Laws prohibiting people with seizures from driving are designed to protect both you and other people from injury. Talk with a licensed healthcare professional to explore alternative transportation methods, join a carpool and offer to pay extra for the gas, contact your city’s public transportation center, ask if your church or a faith-based community organization offers volunteer drivers, or check to see if your community offers a shuttle bus/discounted taxi services for seniors or those with a disability.

Chapter 8: Pediatric Brain Tumors

Children are not smaller versions of adults. Their bodies and brains are still developing, their needs are different, and their tumors are different too. The most common childhood brain tumors are not the same as the most common adult brain tumors.

Though rare, brain tumors are the most common form of solid tumors among children under the age of 15 and represent about 20 percent of all childhood cancers. Childhood tumors frequently appear in different locations and behave differently than brain tumors in adults. Treatment options vary and can be strongly influenced by the age of the child. Children with tumors may also have a much better prognosis than adults with a similar condition.

In this chapter, you will learn more about the characteristics of brain and spine tumors, as well as the most common forms among children.

BRAIN TUMORS IN CHILDREN

Most pediatric brain and spine tumors are primary tumors, meaning they originated in the brain or spine. The most common types of brain tumors in children are astrocytoma, medulloblastoma and ependymoma (see Chapter 3 for specific tumor descriptions). This is a listing of pediatric tumor types:

- > Astrocytoma (pilocytic astrocytoma, high grade astrocytoma, optic pathway low grade glioma)
- > Atypical teratoid rhabdoid tumor (ATRT)
- > Brain stem glioma
- > Choroid plexus tumors, choroid plexus carcinoma, choroid plexus papilloma
- > Craniopharyngioma
- > Cysts
- > Desmoplastic infantile astrocytoma
- > Ependymoma
- > Ganglioglioma
- > Germ cell tumors
- > Glioneuronal Tumor
- > Medulloblastoma
- > Neurofibromatosis associated tumors including optic pathway low grade gliomas
- > Oligodendroglioma
- > Supratentorial PNET
- > Spinal cord tumors

If your child has been diagnosed with a brain tumor, you probably have very specific questions. Your doctor can respond best to your concerns about the tumor, the treatment plan and what to expect in the future.

THE CARE TEAM

Pediatric brain tumors require specialized care that is most often provided by a team of specialists at a large medical center or pediatric hospital. The team may include:

- > Pediatric neurosurgeons
- > Pediatric radiation oncologists
- > Pathologists
- > Endocrinologists
- > Neurologist
- > Genetic counselors
- > Rehabilitation specialists in occupational therapy, physical therapy and speech therapy
- > Pediatric nurse specialists
- > Social workers
- > Child life specialists and others

Additionally, since most of these programs are at teaching hospitals, medical residents and fellows may be involved in your child's care.

Parents or other significant caregivers are also critical members of the care team. You are your child's best and most important advocate when it comes to medical care. You will have lots of questions. You will have to make many decisions. Talk with your medical team early and often. It can take some time for all of the pieces to fall into place. Be patient with yourself. There can be a good deal to learn.

TREATMENT OPTIONS

Treating brain and spine tumors in children is different than treating adults. Children's brains and bodies are still developing, so there are different considerations and standards of care. For most children, treatment starts with surgery. A pathologist will analyze the tissue obtained to classify and grade the tumor.

Following surgery, additional treatment may be required. Possible therapies include:

- > Chemotherapy
- > Conventional radiation therapy
- > Proton Beam radiation
- > Stereotactic radiosurgery, a precise form of radiation therapy
- > Stem-cell rescue, blood and marrow transplantation
- > Interventions to address side effects of the tumor or the treatment
- > Rehabilitation to regain lost strength and skills
- > Ongoing follow-up care for long-term management and to detect recurrence of the tumor

Clinical Trials

Your doctors may suggest that your child be enrolled in a clinical trial. Clinical trials offer patients access to experimental therapies that may not be available elsewhere. Talk with your doctor to better understand the risks and benefits of clinical trials.

MAINTAIN YOUR CHILD'S MEDICAL INFORMATION

It is important that you keep records of your child's treatment. Survivors of childhood brain tumors need to be monitored throughout their lives by doctors who are aware of their medical history. As your child matures into adulthood, his or her future doctors will need to know the history of the tumor, completed treatments, specific procedures and drugs used. Sometimes survivors of childhood brain tumors experience side effects later on in life from the treatments they have received. These are called late effects. Access to detailed medical information will help your child's medical team monitor and address any late effects of treatment.

IMPACT OF A PEDIATRIC BRAIN TUMOR ON THE FAMILY

When a child has a brain tumor, it affects the entire family. Emotions can be raw. Stress and anxiety levels rise. Family schedules are more hectic than ever. Parents and others can quickly find themselves sleep deprived, worried and stretched beyond their capacity.

To help you, the ABTA offers information and resources for parents, caregivers, patients and others. To learn more about how this diagnosis can affect your family and learn ways to help cope with the challenges see Chapter 9 on Coping.

TALKING TO YOUR CHILD OR TEEN

A child or teen diagnosed with a nervous system tumor may have many questions. You do not have to have all the answers when your child is the patient, but you do need to have conversations. Most medical teams have a social worker or child life specialist who can help you explain the diagnosis, tests and treatments in a way your child or teen will understand. Accurate information presented in a non-threatening, age-appropriate way can prevent children from imagining explanations that are scarier than the truth and can help them better cope with tests and treatments. Avoiding the topic does not eliminate the concerns or questions your child has, even if they do not express them.

Open, honest and loving conversations help to address fears and misconceptions. Create an atmosphere where your child will want to ask questions or return to the topic whenever new questions or concerns arise. Some children will want lots of details, others will not. Follow your child's lead.

Discussing the diagnosis, tests and treatments is also an important opportunity to reassure your child or teen. Often times, children and teens have misconceptions that must be addressed. The following are responses to some of these misconceptions:

- > This illness is not the result of something you did or did not do. It is not related to your behavior or your thoughts. It is not a punishment.
- > A tumor is not contagious. You cannot “get it” or “give it” to anyone.
- > You are still you. Having a tumor in your brain does not change who you are. It also does not change my love for you.

Be prepared for difficult questions including “Am I going to die?” Be honest and direct. Tell your child that everyone is different and reacts differently to the illness and the treatment. Together with your medical team, you are doing everything you can to make the illness go away.

For more topics or sample explanations for talking to children with a brain tumor, see Chapter 9.

SIBLINGS

When a child has a tumor, the impact of the situation quickly extends to the entire family. Routines are disrupted. Priorities are shuffled. Parents are pulled away from other day-to-day activities to attend to the needs of the child. Necessity may require that brothers and sisters be left in the care of other family or friends.

Siblings can feel confused, neglected and guilty. In the rush of medical needs, it can be easy to short change conversations with brothers and sisters.

Even at an early age, children sense when there is something wrong. They may even think that they created the problem. Keeping brothers and sisters informed helps them feel connected to their sibling. Provide honest, age-sensitive information.

Address Their Fears and Concerns

Common misconceptions arise among siblings too. “Can I catch it? Did I cause it? Will I get it?” The social worker or child life specialist on your medical team can also help you communicate effectively with family members.

Engage Them

The treatment process can be lengthy. Siblings can feel as if all the attention and energy is spent on just one child in the family. Once the immediate crisis passes, find ways to reconnect. Share a meal, a goodnight book, a conversation while driving. Focus on quality, not quantity. Even if you have just a little time to spare, devoting your undivided attention to others in the family will make a difference.

Involve Them

Oftentimes, brothers and sisters want to help. If this is the case, try to find ways for them to help. It will give them a concrete way to process their own emotions and demonstrate some small measure of control over what is happening in their lives.

IMPACT ON RELATIONSHIPS

Although a crisis can sometimes bring out the best in relationships, that is not always the case. A crisis and its aftermath can strain healthy relationships and expose cracks in struggling relationships.

Whether you are married or divorced, maintaining a relationship with a co-parent can be difficult while caring for a child with a brain tumor. Issues you have struggled with in the past can return.

Guilt, anxiety, exhaustion and stress can complicate relationships even further. Try to maintain communication and honor the experience of others going through the situation. Be open and honest with your care team regarding relevant family history, custody and other emotional concerns that may spill over into the care environment.

Be aware of the problems that can emerge in other family relationships, including those with extended family members, in-laws, step families, etc. Each person has a legitimate interest in the child and the child's treatment. Yet, it can be very frustrating and time consuming to communicate with each of them. Consider designating a close friend or other family member to serve as a "spokesperson" to keep all informed and updated.

PRESENTING/ACUTE SYMPTOMS

Patients may present with disability at diagnosis or neurologic problems may worsen with surgery, but often with supportive treatment, many of the deficits improve or resolve with time.

A majority of pediatric brain tumors are located in a midline location in the brain, which may result in hydrocephalus (buildup of fluid). Sometimes removing the tumor resolves hydrocephalus, but many kids may require an additional procedure to manage hydrocephalus (Endoscopic Third Ventriculostomy or Shunt placement).

LATE EFFECTS OF TREATMENT

People who are treated for a nervous system tumor may experience side effects long after treatment has stopped. These complications are known as late effects of treatment. Late effects are not unique to children. However, because children can live for many decades post-treatment, it is essential that parents and other caregivers be aware of these effects.

Late effects vary considerably based upon multiple factors, including a person's age, tumor type and location, and treatment type and duration. Surgery, radiation therapy and chemotherapy can all contribute to late effect complications. It is extremely important that brain tumor survivors of all ages be followed by a medical team versed in late effects throughout their lifespan.

Some of the more common complications of treatment are listed below. Some can be addressed through rehabilitation and other accommodations. All require ongoing monitoring.

- > Physical disabilities, such as weakness of muscles and diminished coordination
- > Learning disabilities, including problems with memory, attention, comprehension and information processing
- > Behavioral changes and emotional issues
- > Hearing and vision problems
- > Issues with hormones (endocrinopathies)
- > Social integration issues
- > Seizures and other neurological issues
- > Hormonal problems, including slowed growth, hypo- or hyperthyroidism, diabetes, early or late puberty, and infertility
- > Damage to internal organs or other body systems
- > The possibility of developing secondary cancers in other parts of the body or a recurrence of a tumor in the brain

Lifetime Monitoring & Maintaining Medical Records

Childhood brain tumor survivors need to be closely monitored. Most large pediatric brain tumor treatment programs have specific survivor programs. These programs provide specialized health monitoring throughout childhood. They also identify specific risks based upon a thorough review of the child's medical history, including identification of risks associated with specific tumor types or interventions. As a child enters adulthood, survivor programs help families transfer ongoing care out of the pediatric setting and into a medical program that can provide the specialized monitoring the patient will need as an adult.

Parents and caregivers should maintain or know how to access a complete, detailed medical record of their child's treatments. As more information is known regarding the effects of specific treatments, your child's medical record will provide the information needed to better understand any risks and influence the recommended course of follow-up.

Your medical team is the first line of defense to address late effects and to ensure close monitoring of your child's condition.

RETURNING TO SCHOOL

"I think it's time to go back to school." After weeks or even months of treatment and recovery, these words are cause for celebration among children and parents. For many, returning to school is an important milestone on the road to recovery.

As wonderful as it is, the transition to school does require special attention and care. Your school-age son or daughter may have significantly different needs today, and you will probably have many concerns and questions. Additionally, for some, a return to school is not a singular event as much as it is a fluid shifting of time spent at home, at school and in care.

Start Early

Returning to school is an exciting time, but it can be overwhelming for parents and children. Returning students have weathered difficult treatments that may have altered learning capabilities, behavior, strength, energy levels, coordination, speech, hearing or eyesight. It may be the first time your school has worked with a family in your specific situation. Communicating with the school early and often during treatment will help smooth the way for a good transition.

While your child is still recovering in the hospital, update their teachers regarding diagnosis and treatment. It is critical to talk to your child about what he or she may (or may not) be comfortable revealing about this personal situation. Once informed, teachers and classmates can be a tremendous source of much-needed support during hospitalizations and home stays through cards, letters, phone calls, texts, social media and personal visits.

Try to maintain education goals during treatment. Work with the school to complete assignments at home during recovery. Read to your child and keep them engaged with learning as much as possible during the time away from school.

Tap Into Special Education Services

As a return to school nears, meet with the principal and primary teacher and/or special education coordinator to discuss necessary accommodations. Bring brain tumor information from the ABTA with you to share. This is your opportunity to discuss your child's diagnosis, treatment, and the resulting physical, neuropsychological, emotional, social or behavioral changes.

If your child has physical or learning disabilities following a brain tumor diagnosis or related treatment, your child may qualify for benefits or accommodations under one of two federal mandates: The Individuals with Disabilities Education Act (IDEA) or The Rehabilitation Act of 1973 – Section 504.

To access services under these mandates, request a school evaluation for your child. This will include a series of educational tests to determine how your child learns best and what type of accommodations will help your child to optimally learn. These services are usually coordinated by your school district's special education department.

Neuropsychological Testing

Children treated for brain tumors may experience neuropsychological effects following treatment. Neuropsychological testing is done to help define the impact of a tumor and its treatments upon the child's ability to learn. It can be used to assess processing speed, attention, visual motor integration, planning and organizing skills, visual and verbal memory, reading comprehension, math calculation, and applied abilities.

Ideally, baseline testing is done before treatment starts and is done again prior to returning to school. Neuropsychological testing will help assess needs and identify the necessary accommodations for a student to succeed in a classroom environment.

The neuropsychological testing may be used to complement school administered evaluations.

Monitor and Advocate

When your child first returns to school, you may wish to plan regular meetings with their teacher or counselor to better understand how things are progressing in the classroom. Ask about your child's behavior, signs of fatigue, excessive frustration or depression. Check in frequently with your child. Ask questions. Monitor performance. Be aware of changes and be prepared to request modified or new accommodations at school as the circumstances require.

You must advocate for your child's education in the same way that you advocated for medical care. Work closely with teachers and the school. Involve your medical team if necessary.

For more information on pediatric brain tumors, please visit www.abta.org, call our CareLine at 800-886-ABTA (2282) or send us an email at abta cares@abta.org.

Chapter 9: Coping

Nothing can prepare you for a brain tumor diagnosis. Whether you or a loved one is diagnosed, it is something that takes time to process. You may be experiencing feelings of fear, uncertainty or isolation.

It is important to know that these feelings are normal and that you are not alone.

This chapter offers some helpful suggestions from professionals who specialize in helping people cope, as well as some practical advice from other brain tumor survivors and their loved ones.

YOUR FEELINGS AND REACTING TO THE DIAGNOSIS

A brain tumor diagnosis can cause many emotional reactions: shock, denial, anger, resentment, guilt, reflection, anxiety, loneliness, depression, resignation, acceptance and hope.

Along with an initial shock, many people go through a state of temporary denial. Patients and family members may feel traumatized and depressed, may become numb and hide, or may deny their feelings. Some people may refuse to discuss or even acknowledge their diagnosis. Denial can last a few days to a few weeks or even a few months.

As time passes, denial often evolves into a feeling of anger; many patients question, “Why me?” Life may feel unfair. Some develop a sense of resignation about living with the physical or emotional changes caused by a brain tumor diagnosis. Others feel resentment because they are impacted by this disease and other people are not. For some people, this resentment may turn into depression.

Over time, a sense of acceptance will set in as you start to realize the brain tumor is a reality. When you come to grips with the diagnosis, you can begin to plan how to move forward. This may be a time to reassess your values, think about how you want to live your life and look at your career choices. Find ways to maintain positive, close and loving relationships with your family and other important people in your life.

Although this may be hard to believe (especially for newly diagnosed patients), some people feel that their personal lives change for the better after a major diagnosis, such as a brain tumor. A brain tumor diagnosis makes people re-evaluate and change their lives, often in a much more positive direction. If you use every minute of the day in a valuable and meaningful manner, there is much to hope for and look forward to.

There is no one single way to deal with your emotions. One day you may feel stable and then feel uneasy again the next day. Not everyone shows their emotions, nor does everyone experience the same feelings. We all have a wide array of emotions and our experience depends on how we acknowledge and cope with them.

A Special Note on Anxiety and Depression

While being treated for a brain tumor, many patients experience anxiety or depression. These two common side effects should also be monitored and treated by your healthcare team to the same degree as any other physical condition.

Anxiety

People often experience anxiety while going through stressful times. Many people feel “anxious” waiting for test results or returning to the doctor for follow-up visits. Symptoms of anxiety include a sense of fear, a feeling that “something bad” is going to happen, a rapid heart rate, perspiration, nausea, shortness of breath, dizziness or a feeling of unreality. Make sure to talk to your doctor about your physical symptoms, even though they may be psychologically based. Sometimes, just the reassurance that your doctor provides will be enough to relieve your anxiety. If your doctor determines that the symptoms warrant treatment, he or she may suggest medication or an appointment with a psychiatrist, psychologist, or social worker.

Depression

Depression is different than just feeling sad or upset. Some of the symptoms of depression are:

- > Sad or depressed for most of the day on most days, or no feelings at all
- > Irritability
- > Loss of interest or pleasure in activities that used to be pleasurable

- > Weight loss or weight gain
- > Sleeping too much or too little
- > Fatigue or low energy
- > Feelings of guilt low self-worth
- > Thinking about death, dying, harming yourself, or suicide

Many of these symptoms are common reactions to nervous system tumor treatment.

If these feelings persist for more than two weeks or if they are severe, talk to your doctor. Your doctor will determine whether these are signs of major depression and, if so, will provide direction. The doctor may prescribe medication or suggest a psychiatric consultation. Depression is treatable, but must be diagnosed first.

INCLUDING FAMILY AND FRIENDS

Life is going to change. For many, sharing their situation with family and friends can offer extra support. However, telling your family and friends that you have a brain tumor can be difficult. If you are uncomfortable with doing this on your own, consider finding a care conference or scheduling a meeting with your doctor, healthcare team, and the primary members of your family. Written publications and educational materials about brain tumors can serve as a supplement to the conversation by providing helpful, easy-to-understand information.

Like you, your family needs time to process and understand your diagnosis. Your family will be more supportive and helpful if they are able to completely understand your diagnosis and available treatment options.

Social workers can help with communication challenges between you, your family, and friends by facilitating conversations about associated thoughts and feelings. A social worker can also suggest appropriate coping techniques. To locate a social worker, try contacting the social services department at your local hospital. In addition, social workers are available at community centers, social service agencies, government health agencies and schools.

If friends offer to help, accept their offers. You will benefit from the assistance, and your friends will feel needed. Groceries, laundry, driving, weeding the garden, providing meals on days of your doctor visits, driving you to the clinic for therapy – there are many possibilities. Keep a “wish list” of things you wish you had the time to do. When someone offers to help, pull ideas from that list. Or create an online calendar with tasks that need to be done.

Although many people will be supportive, there will be some who simply find it difficult to deal with or even acknowledge your illness. Some people may avoid contact or conversation because they do not know what to say. Also, be prepared for well-meaning friends and neighbors who insist upon telling you stories about “miraculous” cures. Do not let their second- and third-hand news make you feel obligated to start yet another information search. Thank them for their concern, but remember that what works for one person may not be appropriate for another. There are many different types of brain tumors and many different treatments. If you have questions, ask your doctor.

CHANGES WITHIN THE FAMILY

With time, a circle of friends will emerge that you are comfortable discussing what you are going through with and that you can count on for help and support.

Shifts may occur within the family system as everyone reacts to the brain tumor diagnosis. A spouse or significant other may begin to worry more or wonder how things will get done. Spouses

or significant others may feel a sense of loss as they assume the role of caregiver or may seem bewildered as they take on responsibilities previously handled by the other person.

Every marriage has “unwritten” rules that define how the relationship and the family functions every day. Over time, spouses take on certain roles, like being the caretaker or the provider for the family. They work out the labor issues for the household and establish guidelines for their intimate relationship and personal friendships. However, the diagnosis of a brain tumor can change these rules. Spouses may have a hard time adjusting, especially if the caretaker and provider roles have changed. Some couples find they start feeling uncomfortable with each other, they begin to feel distant and that they are losing the intimacy of their relationship. You may begin arguing or disagreeing with each other. During moments like this, it may be tempting or seem easier to pull further apart. Instead, make a dedicated effort to talk with your spouse about the changes that are taking place and the emotions involved.

While the news of a brain tumor diagnosis will initially disrupt your personal and family life, when you come to terms with the news, you may find it is an opportunity for renewal, hope and a new relationship. Talk with your spouse about the changes in your lives. Acknowledge the need for flexibility now, and find some rules that you can both agree upon and live by. If you have a difficult time doing this, seek the advice of a trusted friend, a religious advisor, a social worker or a psychologist. A professional may be able to help with the adjustments and changes associated with the medical diagnosis and its effect on your relationships.

Think about the ways you have handled a previous crisis in your life. Are you comfortable sharing your concerns? If so, whom do you usually turn for support? Do you prefer sharing with a close family member or a close friend, or do you turn to those you may not know well, such as a counselor or a support group? Do you find comfort in religion? Think about the methods you normally use to deal with problems. Even a long shower, relaxing music or meditation techniques can be of help.

As you move along through your brain tumor journey, you will begin to find resources you did not know existed. There are people willing to help – you are not alone. Reach out, learn as much as you can and become an active participant in your health care. Begin by eating healthy meals, even when you are not very hungry. Moderate exercise (if approved by your doctor) can help fight fatigue, depression and improve your sleep. Remember that it is normal to feel unhappy sometimes, but realize there is help available to those whose sadness is consuming or excessive.

TALKING WITH CHILDREN

Before talking with your children about your (or your spouse’s) brain tumor diagnosis, try to anticipate your children’s reactions and concerns. Children use their imaginations to fill in the gaps; their fantasies can cause undue fears and anxieties. Give children information in words they understand. Use their questions as a guide to the amount of information they want; do not provide them with more than they ask. Be prepared for questions that are not easy to answer and reply honestly and simply in words appropriate for their age.

Remember that very young children have little experience with disease – their first questions may focus on the practical. They may want to know who will prepare dinner or put them to bed. Tell them what it means to “go to” or “be in” the hospital. Simple drawings may help. Use their questions as a guide to the information you provide to them.

There are many books available that can help parents explain their illness to children. Read these books with your children and offer them the opportunity to ask questions or express their fears and concerns. Young people often have remarkable insight and can be a source of great comfort. Most importantly, remember that children of all ages need to be reassured that you have planned for their needs. Explain those plans and arrangements to your children, making sure they know you are still very much involved, even if from a distance.

How Do I Tell Them? What Do I Say?

These sample explanations can be adapted for conversations with children.

- > “The doctor wants to do some tests to find out why I am getting sick to my stomach and having headaches...”
- > “A neurosurgeon is a doctor who knows a lot about the brain.”
- > “An MRI scan takes a picture of the brain, but it cannot see what you are thinking.”
- > “A brain tumor is a lump in the brain that doesn’t belong there. The doctor is going to take it out to help get rid of the headaches.”
- > “A brain tumor is a collection of abnormal ‘cells’ in the brain that are growing out of control. These cells were originally normal brain cells, but something inside of them changed. For no good reason, they started to divide and make more of themselves. This growing collection of abnormal cells is called a tumor.”
- > “With a tumor in there, the computer center of my brain can’t work the way it is supposed to. That’s why I have headaches and seizures.”
- > “No one knows for sure what causes a brain tumor. They just happen. But we do know that nothing you did or thought or said caused the tumor. Nothing you ever wished made this happen. Nothing your brother or sister or friends said made this happen. We also know that you don’t ‘catch’ brain tumors from other people.”
- > “Would you like to talk about this? Is there anything that you would like to ask?”

Above all, reassure your children they are loved and will be taken care of.

Additionally, try to ensure that routines within the family change as little as possible. Children find the “daily rules of family life” very important. They feel safe and thrive under day-to-day conditions that are consistent and easy to predict. Therefore, make sure you and your spouse agree on what to tell your children, and how to keep their lives as normal as possible. If schedule changes become necessary, try to make the new schedule repeatable so it becomes a routine. The sameness will be a comfort to your children.

Older children may link the diagnosis to stories of illness they have heard from friends or to other personal experiences with grandparents or neighbors. Give older children the opportunity to ask questions and share their concerns. Saying, “Is there anything you’d like to ask?” can be difficult, and children may be reluctant to answer the question. Let them know that you want to have an open conversation. This can help clarify misinformation, as well as provide reassurance.

Some children, especially pre-teens and teenagers, may begin to act out. They may start getting in trouble, begin having problems in school or start doing things they normally would not do. This is usually a sign that the child is having trouble coming to terms with what is happening within the family, and that more direct action is necessary. A serious discussion with the child is very important at this stage. Involve them in the day-to-day family activities. If the behavior persists, seek professional help, such as counseling or family therapy. Many families find that after the initial diagnosis children learn to adjust and adapt to the news, especially if the parents are consistent and in agreement regarding family matters.

Most importantly, remember that children of all ages need to be reassured they are loved. They need to know that they will be cared for during the parent’s illness. Neighbors and relatives can help keep the children’s routines as normal as possible. Plan ahead for special events. If an ill parent is unable to make the school play or watch softball practice, perhaps an aunt, uncle or

friend can fill in. Although it will not be the same, the child will know that mom or dad cared enough to ensure that someone would be there.

REDUCING STRESS

For most people, fears of the unknown and uncertainty for the future can cause great stress. This is normal. Give yourself permission to be temporarily overwhelmed. Then, take a few deep breaths and begin to think about the things you can control. Here are a few ideas for reducing stress:

- > Ask family and friends to help with household responsibilities.
- > Find someone to assist you in completing medical forms and claims.
- > Participate in planning your treatment.
- > Help determine your medication or treatment schedules.
- > Decide which chores are important and which can be temporarily ignored.
- > Choose to share or not share your experience with others. The choice is yours.
- > Be kind to yourself by listening to music, journaling, reading a book or taking a mid-afternoon nap – indulge in relaxing activities that can recharge you.

If you are a family member or a caregiver, let yourself to take some “time off” to focus on your own needs. Call upon other relatives or friends to serve as relief workers so you can take a much needed respite.

Communication is an important part of reducing stress. Talk to your family about your needs, feelings and responsibilities. Listen to their concerns as well. Sometimes one person will take on too many responsibilities. Or, in trying to protect others, a family member may not express his or her own needs. Taking the time to talk about what needs to be done and who can reasonably do it allows everyone to feel useful and avoids feelings of resentment. Relaxation, meditation or imagery techniques can also help reduce stress for you and your family. Consider taking a class together.

Birthdays, holidays or anniversaries can be a difficult time for your family. Anxiousness or irritability around these days is normal. Plan ahead and make activities simple and memorable.

If anyone in the family is having difficulty adjusting to the diagnosis, family therapy may be helpful. A trained, professional therapist, especially one specializing in the treatment of chronic illness or grief reactions, can help guide families through this difficult time. Close friends, religious leaders or your healthcare professional can be a source of emotional and physical strength. Friends may be able to search for community and medical resources of value to you. Contact your library, local civic organizations, village hall or religious institutions. Learn what community programs are available in your area and take advantage of their services. Each resource you find makes it easier for you and your family to cope with your new situation.

From Patient to Patient: Suggestions for Coping and Managing Stress

These suggestions were provided by brain tumor patients and their families. We hope you find some ideas to be helpful.

- > Get a second (or third) opinion from doctors who specialize in brain tumors. To be an effective advocate for yourself, you will need information on all your options.
- > Ask your doctors this question: “What questions should I be asking that I do not know I should be asking?”

- > Do not be afraid to talk about your fears and feelings – you are not alone. People do care and are willing to help.
- > Relax your need for control. You will save your energy.
- > Take advantage of help offered to you. Family, friends and neighbors can make life easier, if you let them. And do not worry about repaying the favors.
- > Allow yourself to cry. It is a good escape valve for both women and men.
- > Set short term goals so you can feel good about your progress.
- > Find one thing good about each day.
- > Laughter helps.
- > Find purposeful things to do – a daily morning walk with a friend or invite your grandchild to lunch weekly.
- > Keep a journal. It is a special place all for yourself.
- > Decorate a small box with bits of costume jewelry, brightly colored paint, wonderful pictures from old magazines. Put your worries in it and tightly close the lid.
- > Be kind to yourself. Take time to pamper yourself, make time to play, be lazy when you need to.
- > Set your own limits, not those expected by others.
- > Know that symptoms usually worsen in the darkened evening hours. This is generally when everyone is tired and defenseless and at their worst. Arranging for outside support or visitors in the evening may help.
- > Find someone to confide in. For both patients and families, support groups can be invaluable. They can help you know you are not the only one dealing with this situation.
- > Take breaks from the brain tumor world. Go to a movie, have a picnic in the park, invite friends to join you at a concert. Make time to do this regularly, even if “regularly” is only once a month.
- > Find a spiritual leader or contact your clergy.
- > If you are having a difficult time coping, seek professional help.
- > Read about brain tumors, new treatments and about brain tumor survivors. Be positive and do not feel defeated.

REACHING OUT FOR SUPPORT

Most of us do not want to be alone when facing a crisis. Emotional support from family, friends and loved ones can give us comfort and strength. However, this support may not be enough and you may feel a need to connect with others going through similar situations. There are many ways to reach for support but not all types of support work for everyone.

Support Groups

Patients and families often find help through brain tumor support groups. A support group is a face-to-face gathering of people seeking to share their experiences with the help of a support group facilitator. They come for emotional, social, and possibly spiritual support. There are different types of support groups for adults, parents of children with brain tumors, children and siblings. While not for everyone, support groups can provide a comfortable, safe place to share

experiences and concerns, as well as practical information and strategies for dealing with the diagnosis and treatment of a brain tumor.

Support groups are located throughout the country, and most are open to both patients and family members. In large groups, patients and family members may meet separately. Support groups often vary in their formats. Some groups have an open discussion format, while others are more educational. Other groups offer a little of both.

Groups may meet weekly, monthly or quarterly, depending on the needs and desires of the group members and the availability of the facilitator(s). Some groups are professionally facilitated and some are not. In a professionally facilitated group, a social worker and/or nurse provide guidance and direction. They also monitor member interaction to ensure that everyone's needs are being addressed. Groups that are not professionally facilitated may be less structured and more closely resemble a self-help group.

Many times, face-to-face support groups provide a closeness and warmth among the participants who interact with and support each other on a very personal level. However, face-to-face support groups are not a viable option if there is not one in your community and/or if the group location and meeting time are not convenient for you.

If you are not comfortable with a particular group or it does not meet your needs, try another one. Finding the "perfect" support group can take time and can be a process of trial and error.

To find a support group near you, visit www.abta.org, call 800-886-ABTA, or email abta-cares@abta.org.

Online Support Resources

For people who do not have access to a regularly scheduled face-to-face support group or for those who prefer email communication, online support resources are an option. There are many different types of online forums that allow you to communicate with one or many individuals who share your specific circumstances.

Chat rooms are generally real-time discussion forums, similar to a phone call among a group of individuals, although the conversation is written and not heard. These discussions may or may not be moderated. In a moderated discussion, someone is screening the content to ensure that only appropriate text is posted.

Email discussion groups, sometimes called "listservs," and bulletin boards are forums where messages can be posted and read by individual subscribers at their convenience. This form of communication is more like reading a letter than talking on the phone. The letter can be read, thought about and responded to at any time. Most listservs require that users subscribe or register prior to sending a message. Some are moderated, some are not. In moderated groups, someone is responsible for screening the messages to ensure that they are not offensive, and that they pertain and conform to the specified subject and purpose of the forum.

Another factor to consider is the privacy policy of an online forum. A privacy policy lets users know whether or not their personal information is being collected as they use the forum, how that information is stored, and whether or not it is shared with people outside the forum.

Online forums allow individuals to communicate with a potentially large number of people, far beyond those in an immediate geographic vicinity. Additionally, participants can go online to "check-in" with the forum, read and send emails at their leisure.

On the downside, in addition to the privacy concerns and the potential anonymity of participants, online communications – while seemingly with one person or a small group – are available to the entire site or forum membership. This may include "lurkers" – people who are anonymously and invisibly monitoring discussions. Despite the best efforts of sponsors to adhere to their

privacy policies, Internet communication is never private. Therefore, with a large group of people unknown to you, it is best to only share information that you are completely comfortable sharing with strangers.

ABTA Connections Online Support Community

The American Brain Tumor Association's Connections online support community connects patients, families, friends, and caregivers for support and inspiration. Unlike other social media outlets, Connections is a more private setting where members can ask questions of their peers, provide updates on their personal situations, and gain confidence in and comfort through communicating with others who may be going through a similar brain tumor journey. Members also maintain full control of their privacy settings without the distraction of ads and instant messages. Learn more at www.abta.inspire.com

Individual/Family Counseling

Professional counseling can help with more complex issues or when informal support does not adequately meet someone's needs. Clinical social workers, clinical psychologists, marriage and family therapists, and clinical professional counselors can offer one-on-one and family counseling when more formal assistance is needed.

Neuropsychologist

A neuropsychologist is a mental health professional with expertise in assessing and treating problems of psychological function and behavior relating to the brain and central nervous system.

A neuropsychological evaluation involves a variety of tests to assess one's ability to remember, pay attention, use language, problem-solve and conceptualize. It also can assess perceptual and motor abilities, emotional state, behavior, and personality. A complete assessment can take six to eight hours.

Neuropsychological treatments include teaching ways to help individuals improve their impaired functions by learning strategies to compensate for lost or weakened abilities, despite limitations.

When seeking a neuropsychologist, one factor to consider is whether or not the psychologist is board certified. Board certification refers to the certificate received once post-training examinations have been successfully passed. This is a mark of high distinction in the profession.

WHERE DO WE GO FROM HERE?

Eventually, the treatments are completed and the medical appointments get further apart. The pace slows and another period of adjustment begins. During this time, it can be difficult to do nothing after having done so much.

Your task is to become well again. Make appointments for your follow-up doctor visits or scans and mark them on your calendar. Begin to rebuild your life within the guidelines set by your healthcare team. Learn about healthy eating. Exercise within the guidelines given to you by your doctor. Get out, see friends and be good to yourself. But be patient; getting well takes time.

The brain tumor world will always be some part of your life. Learn where it belongs and what feels comfortable for your family. Your priorities will also change with time. That is okay. The changes reflect your ability to adapt. Talk with your family and build this new future together.

You and your family may continue to have questions about living with a brain tumor, wish to keep abreast of the newest findings in brain tumor treatment or learn more about the research that will someday lead to a cure. The ABTA's website – www.abta.org – offers extensive brain tumor information, treatment, and research updates, as well as information about upcoming ABTA meetings and events.

Chapter 10: For Caregivers

When someone you love is ill, naturally you want to do everything you can to help them. Being a caregiver to a family member or a close friend can be one of the most rewarding experiences a person can have. However, it can also be frustrating, lonely and overwhelming, especially when combined with the ongoing stresses of everyday life. Remember that you are not alone and resources are available to help you as a caregiver.

Trying to manage the logistics – not to mention the emotional impact – of a brain tumor diagnosis can challenge even the most caring and compassionate caregivers. Taking care of yourself is just as important as taking care of your loved one. This includes being kind to yourself, asking others to help, taking time away from the patient, or allowing yourself a quiet moment to think and reflect. The following suggestions can help you get the assistance you need while ensuring that you continue to care for yourself. As with any list of suggestions, pick those that work best for you.

ALLOW YOURSELF TO ASK QUESTIONS

Hearing the words “brain tumor” for the first time can be overwhelming. It is common for families to make a visit to the doctor, hear terms and phrases they have never heard before, and then be asked to make a decision. Once you get home, you may begin to question what you heard and if you properly understood any of it. If you have questions about the information your family was given, if you did not understand something that was told to you or if you have additional questions you forgot to ask, call the doctor. Unless you speak up, the doctor will assume that you understand everything. Having answers to your questions can be a great stress reducer.

ASSESS WHAT YOU NEED

Have a clear sense of what you need in order to be a successful caregiver. To do this, ask yourself: “What help or information, if it were available, would make caregiving significantly easier right now?” Be very specific. Do you need help with grocery shopping or need someone to come over a few times a week so you can get out of the house? If you need to talk to someone, are you looking for a sympathetic ear or someone to help you solve a problem? Be selective about the information and resources you choose. Always come back to the question: “What is it I need?” as opposed to “What is available?”

DELEGATE RESPONSIBILITY

Do not do everything yourself if you have others in your family who can help. Sit down and discuss what each member of the household – including children – can do, and develop a schedule of responsibilities. Take into account each person’s ability, maturity and availability. Remember that not everyone can or should be directly involved in caring for the patient; there are other tasks that also need attention. Look beyond your immediate relatives for help; even those further away can participate on some level. Finding others to handle family tasks will give you more time to care for your loved one and yourself which will ultimately help reduce stress.

ACCEPT ASSISTANCE

If there are no other members of your household or relatives close by, look to friends or members of your church or social group for help. Often, people want to help but are not sure what they can do. Be prepared to respond to their offers. Try to determine the time, money or energy commitment they are willing to make and give them one or two suggestions that fit their level of commitment. Some people may be available on a one-time basis to run an errand, babysit or help with a particular household chore. Others may be available more regularly or for longer periods of time. Ask for help with grocery shopping, cooking and freezing meals, yard work, household repairs, driving car pool, or driving to therapy appointments. Those living further away may be able to help with financial needs, filing insurance claims or searching for support resources.

If people offer help at a time when you really do not need it, tell them how much you appreciate their offer and their friendship. Suggest they ask again in a few weeks or ask if you may call on them if your needs change. You can also suggest they help in a less hands-on and direct way, such as making a visit, saying a prayer or lending a supportive ear when you need it.

ACCESS HOME HEALTH CARE

Home health care agencies and caregivers are available to offer additional support and respite care. Skilled services may include medical care and/or physical, occupational, and speech therapy. Home health aides may help with personal services like bathing, dressing, eating and household chores. Adult day care is also available in many communities.

UTILIZE ONLINE SUPPORT GROUPS AND WEBSITES

There are a variety of Internet-based resources (online support groups, listservs, chat rooms and message boards) where caregivers can share information and provide support to each other. There are also websites focused on the needs of caregivers. These Internet-based forums offer

an alternative for those who do not have access to a face-to-face support group in their area or who prefer to communicate and seek information online. Most online forums require that you subscribe or register prior to sending messages. Each forum has its own guidelines for etiquette rules and procedures.

ABTA Connections Online Support Community

The American Brain Tumor Association's Connections online support community connects patients, families, friends and caregivers for support and inspiration. Learn more at www.abta.inspire.com.

BE KIND TO YOURSELF

If you are feeling particularly self-critical, stop and ask yourself, "What would I say to a friend who was feeling guilty about something he or she did or neglected to do?" or "Would I think less of my friend or would I understand that he or she is doing the best anyone could under the circumstances?" You will probably come to realize that you would have no problem letting your friend off the hook; therefore, you should apply this mentality to yourself. When you fall short of your expectations, be at least as kind to yourself as you would be to a good friend.

BE ASSERTIVE

Learn to say no. Set limits on your time. Be realistic about what you can and cannot be responsible for right now. Consider asking friends to visit during hours that are convenient for you and your loved one. If you find yourself with visitors who sometimes "wear out their welcome," ask if you might use the time during their next visit to run errands.

TAKE CARE OF YOUR BODY

Many caregivers tend to neglect their own health in order to prioritize the needs of the person they are caring for. However, neglecting your health can leave you vulnerable to exhaustion and disease. To prevent this from happening, do a regular self-check to see how you are feeling. Ask yourself: "How am I feeling today? When was the last time I saw my doctor for a check-up or for treatment of an ongoing problem? Is it time to make an appointment?" Make sure you are eating regular, nutritious meals; exercising (check with a doctor before starting any new exercise routines), and sleeping regularly.

DISCUSS YOUR FEELINGS

Having someone to talk with can also be very helpful. Look for a relative, a friend or a member of your clergy who you feel comfortable speaking with about what you are going through. Let that person know that you do not expect answers or solutions, just a sympathetic ear. If you are seeking advice, look for someone who will continue to be supportive even if you decide to not take their advice. Furthermore, make sure you feel better after talking with the person you select. Support that does not feel like support usually is not.

Support groups with other caregivers are another valuable resource. Many of these groups welcome both family members and caregivers. To find a support group in your area, visit www.abta.org/supportgroups.

If you find that speaking with friends, family and other non-professional support resources are not making you feel better or you continue to feel consistently overwhelmed, consider consulting a mental health professional. You can ask your doctor to refer you to someone or seek out options yourself. Professional mental health organizations and some websites, like psychologytoday.com, can help you find a therapist in your area.

MAKE TIME FOR YOURSELF

Try to take a break from caregiving for a few hours or more each week. Taking time for yourself is not being selfish. While it can be difficult to rationalize leaving your loved one, especially if they

are very ill, time by yourself is critical in order for you to continue caring for your loved one's physical and emotional needs in a loving and helpful way.

Use your time to do something that you enjoy or that you find relaxing. Meditation, guided imagery, and exercise or movement classes can help reduce stress. Even simple everyday activities, such as going for a walk, listening to soothing music or reading a good book can give you a break from your responsibilities and worries.

KEEP IT SIMPLE

Most importantly, try to keep life as simple as possible during this challenging time:

- > **Be flexible.** This is not to discount the value of routines. However, we all encounter situations where we are just “winging it.” Do not be afraid to try doing things in different ways. Re-evaluate your needs and priorities. What was important before your family member became ill, like having a spotless house, may not be important now. On the other hand, some things that were not important before, like getting enough sleep or finding time for you, may be crucial now. Adopt a problem-solving approach. Break down what you need to accomplish into smaller steps and then brainstorm ways to get from one step to the next.
- > **Decide to do one thing today.** This simple strategy can be very helpful when you feel you are not accomplishing anything, despite always feeling busy. Simply decide to do one thing you have been putting off and just do it. It can be something as simple as cleaning out a file, doing the laundry or making a phone call to the insurance company. Once you have done that “one thing,” it is amazing how much better you feel being able to cross it off your to-do list.
- > **Laugh.** The importance of laughter cannot be overemphasized. Laughter can ease tension, promote relaxation and help you reconnect with the joy of living.

THE ABTA IS HERE FOR YOU

You do not have to go through this journey alone. The American Brain Tumor Association is here to help.

Visit us at www.abta.org to find additional brochures, read about research and treatment updates, connect with a support community, or join a local event.

We can also help connect you with additional resources. Contact our CareLine by email at abtacares@abta.org or via our toll-free CareLine at 800-886-ABTA (2282).

Chapter 11: Facts and Statistics

The facts and statistics here include brain and other central nervous system tumors (including spinal cord, pituitary and pineal gland tumors). We update these statistics on our website – www.abta.org – on an annual basis. This publication was last updated in 2018.

These numbers address incidence, survival and prevalence trends and patterns in the United States only. We thank the Central Brain Tumor Registry of the United States (CBTRUS) for their assistance with this update. For more information, please visit www.cbtrus.org

Ostrom QT*, Gittleman H*, Truitt G, Boscia A, Kruchko C, Barnholtz-Sloan JS. CBTRUS Statistical Report: Primary Brain and Other Central Nervous System Tumors Diagnosed in the United States in 2011-2015. *Neuro Oncol.* 2018; 20(suppl 3): v1-v86.

FACTS AND STATISTICS

Brain tumors do not discriminate. Primary brain tumors – those that begin in the brain and tend to stay in the brain – occur in people of all ages, but they are statistically more frequent in children and older adults. Metastatic brain tumors – those that begin as a cancer elsewhere in the body and spread to the brain – are more common in adults than children.

Brain tumors are the:

- > Leading cause of cancer-related deaths in children under age 15
- > Second leading cause of cancer-related deaths in individuals ages 15–39
- > Fourth leading cause of cancer-related deaths in females ages 15–39

Incidence Statistics

An estimated 86,987 new cases of primary brain tumors are estimated to be diagnosed in 2019; this includes both malignant (26,170) and non-malignant (60,800) brain tumors. These estimates are based on an application of age-sex-race-specific incidence rates from the 2017 CBTRUS Statistical Report using 2010-2014 SEER and NPCR data to project 2018 U.S. population estimates for the respective age-sex-race groups.

In 2019, approximately 5,270 children and adolescents between the ages of 0-19 are estimated to be diagnosed with a primary brain or other central nervous system tumor. An estimated 3,720 new cases of childhood (0-14) primary brain and other central nervous system tumors are expected to be diagnosed in 2019.

Prevalence Statistics

A 2010 study estimated that more than 866,096 people in the United States were living with a primary brain or other central nervous system tumor diagnosis. Specifically, more than 138,054 people were living with a malignant tumor, and more than 550,042 people were living with a nonmalignant tumor.

TUMOR-SPECIFIC STATISTICS

- > **Meningiomas** represent approximately 37.1% of all primary brain tumors, making them the most common primary brain tumor.
- > **Gliomas**, a broad term which includes all tumors arising from the gluey or supportive tissue of the brain, represent approximately 26% of all brain tumors and 81% of all malignant tumors.
- > **Glioblastomas** represent approximately 14.7% of all primary brain tumors and 56.6% of all gliomas.
- > **Astrocytomas and glioblastomas** combined represent approximately 75.8% of all gliomas
- > **Nerve sheath tumors** (such as acoustic neuromas) represent about 8.6% of all primary brain tumors.
- > **Pituitary tumors** represent about 16.5% of all primary brain tumors.
- > **Lymphomas** represent 1.9% of all primary brain tumors.
- > **Oligodendrogliomas** represent approximately 0.9% of all primary brain tumors.
- > **Medulloblastomas, ATRT, PNET and all other embryonal tumors** combined represent 0.9% of all primary brain tumors.
- > The majority of primary brain and other central nervous system tumors are located within the meninges (36%), followed by those located within the pituitary and craniopharyngeal duct, cranial nerves, and the frontal, temporal, parietal, and occipital lobes of the brain (22%).

Metastatic brain tumors are the most common brain tumor. Although statistics for brain metastases are not readily available, it is estimated that there are more metastatic than primary malignant brain tumors per year. Lung cancer and breast cancer are the most common cancers that metastasize to the brain.

Chapter 12: Brain Tumor Terminology

Basal Ganglia

The basal ganglia are masses of nerve cells deep within the cerebral hemispheres (the two halves of the cerebrum, the largest area of the brain).

Brainstem

The brainstem is the bottom-most portion of the brain. It connects the cerebrum (largest area of the brain) with the spinal cord. The midbrain, pons, medulla oblongata and reticular formation are all part of the brain stem.

Cerebellopontine Angle

The cerebellopontine angle is the angle between the pons (part of the brain stem) and the cerebellum (second largest area of the brain).

Cerebellum

The cerebellum is the second largest area of the brain and is located in the back of the head between the cerebrum and the brain stem. It consists of two lateral lobes and a central lobe.

Cerebrospinal Fluid (CSF)

Cerebrospinal fluid is the clear, watery fluid made in the ventricles that bathes and cushions the brain and spinal cord. It circulates through the ventricles and around the surface of the brain.

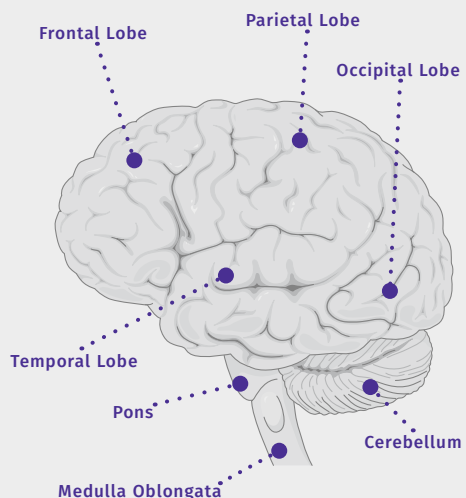
Cerebrum/Cerebral Hemispheres

The cerebrum is the largest area of the brain and consists of the right and left cerebral hemispheres. The right cerebral hemisphere controls the left side of the body and the left cerebral hemisphere controls the right side of the body.

Each hemisphere is comprised of four sections called lobes: frontal, parietal, temporal and occipital. Each lobe controls a specific group of activities.

The outer layer of the cerebrum is made up of gray matter (nerve cells that control brain activity). The inner portion of the cerebrum is mostly white matter with nerve fibers (axons) that are insulated by a fatty substance (myelin). White matter carries information between nerve cells by conducting electrical impulses.

MAJOR PARTS OF THE BRAIN



Choroid Plexus

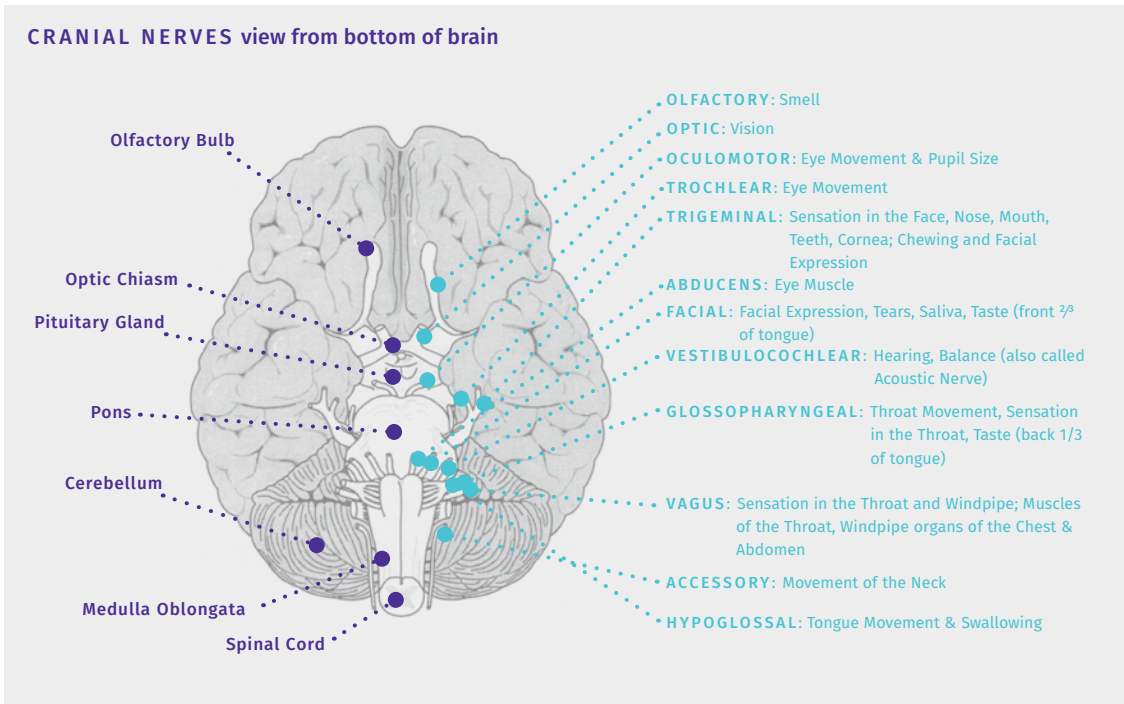
The choroid plexus produces spinal fluid that flows through the ventricles (cavities) and meninges (membranes) surrounding the brain and spinal cord.

Corpus Callosum

The corpus callosum is made of nerve fibers, deep in the brain, that connect the two halves of the cerebral hemispheres.

Cranial Nerves

Cranial nerves are the twelve pairs of nerves that originate in the brain.



Glial Tissue (Neuroglia)

Glial tissues (also called neuroglia or glia) is the supportive tissue in the brain made up of glial cells. The most common glial cells are astrocytes and oligodendrocytes. Ependymal cells are another form of glia.

Glial cells are the origin of the largest percentage of brain tumors, i.e., astrocytomas (including glioblastoma), oligodendrogliomas and ependymomas. Astrocytes are involved with the blood brain barrier and brain metabolism. Oligodendrocytes maintain the myelin covering of nerve cells. Myelin helps transmit information between nerve cells.

Hypothalamus

The hypothalamus regulates sleep cycles, body temperature and other metabolic processes. It acts like an endocrine gland by producing hormones and sending messages that control the hormonal secretions of the pituitary gland.

Medulla Oblongata

The medulla oblongata is a part of the brainstem and connects the brain with the spinal cord. It contains the origins of the 9th, 10th, 11th and 12th cranial nerves.

Meninges

The meninges are three membranes that completely cover the brain and the spinal cord. Spinal fluid flows in the space between two of the membranes.

Meningioma is a tumor that arises from the meninges.

Midbrain

The midbrain is the short portion of the brain stem between the pons (part of the brain stem) and the cerebral hemispheres. The 3rd and 4th cranial nerves originate in the midbrain.

Optic Chiasm

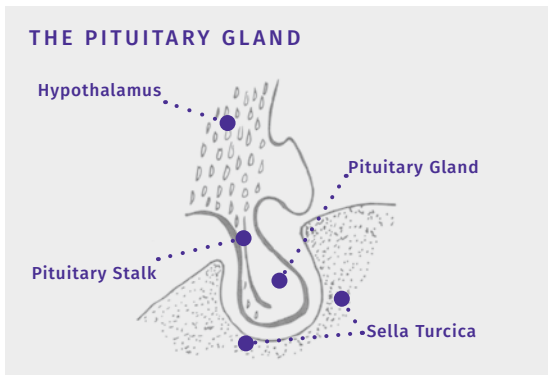
The optic chiasm is the area under the hypothalamus where each of the two optic nerves crosses over to the opposite side, forming an X shape.

Pineal Gland

The pineal gland lies below the corpus callosum (the nerve fibers that connect the two halves of the cerebral hemispheres). It produces the hormone melatonin, which is believed to control the biological rhythm of the body.

Pituitary Gland

The pituitary gland is attached to and receives messages from the hypothalamus. It is composed of two lobes, the anterior and the posterior. The pituitary gland produces several hormones, including prolactin, corticotropin and growth hormone.

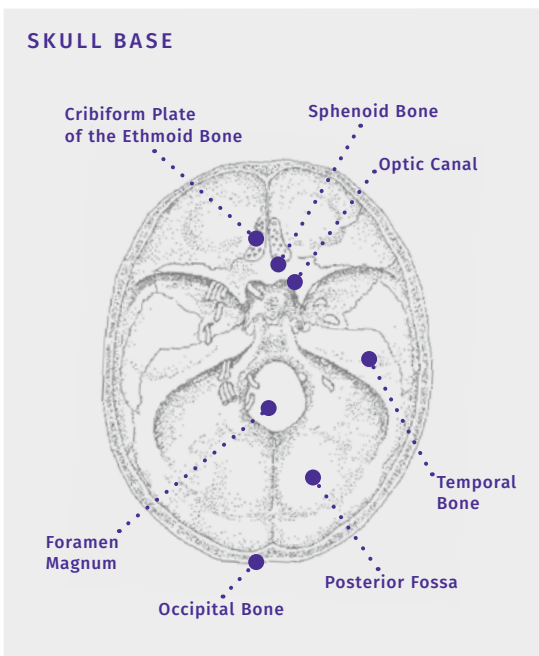


Pons

The pons is a part of the brain stem that contains the origins of the 5th, 6th, 7th and 8th cranial nerves.

Posterior Fossa (Infratentorial Region)

The posterior fossa or the infratentorial region contains the cerebellum and the brain stem.



Reticular Formation

The reticular formation is the central core of the brainstem and controls the level of consciousness. It connects with all parts of the brain and brainstem.

Sellar Region (Suprasellar, Parasellar)

The sellar region is the area around the sella turcica, a hollow in the skull bone that contains the pituitary gland.

Skull Base

The skull base refers to the bony areas that support the bottom of the frontal lobes, the bottom of the temporal lobes, and the brain stem and cerebellum.

Spinal Cord

The spinal cord is made up of neurons and their extensions (nerve fibers). It begins in the medulla oblongata (part of the brain stem) and continues through the hollow center of the vertebrae (the bones of the spine). The spinal cord is covered by the meninges (membranes) through which the cerebrospinal fluid flows.

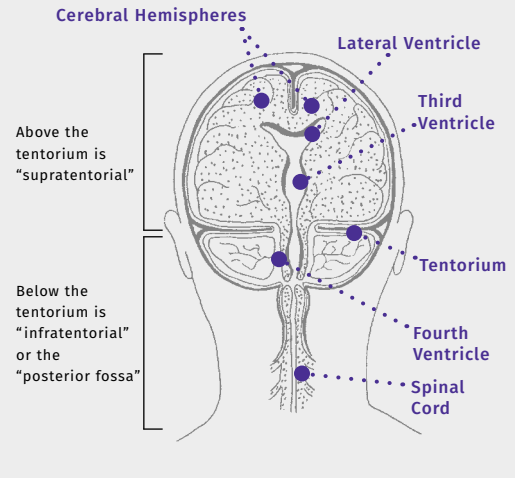
Supratentorial Region

The supratentorial region contains the cerebral hemispheres.

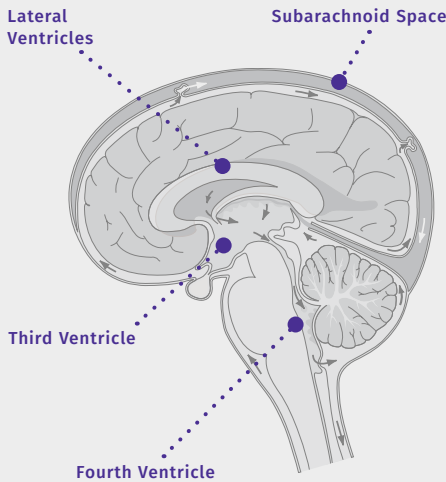
Tentorium

The tentorium is a flap of meninges separating the supratentorial region (cerebral hemispheres) from the posterior fossa (cerebrum and brainstem).

THE TENTORIUM



CSF AND VENTRICLES



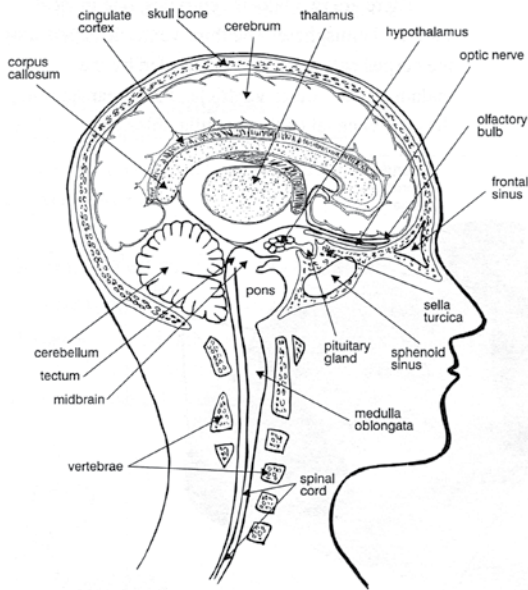
Thalamus

The thalamus surrounds the third ventricle. It processes and relays sensory information and regulates motor functions.

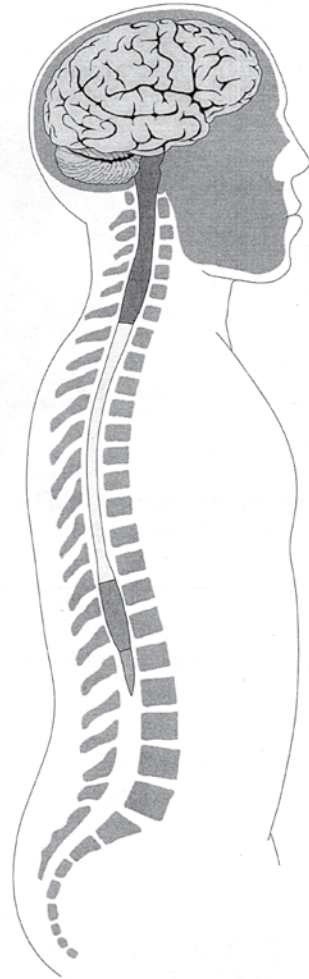
Ventricles

Ventricles are connected cavities that contain cerebrospinal fluid. The fluid is produced by the choroid plexus and flows through the ventricles and the space between membranes of the meninges.

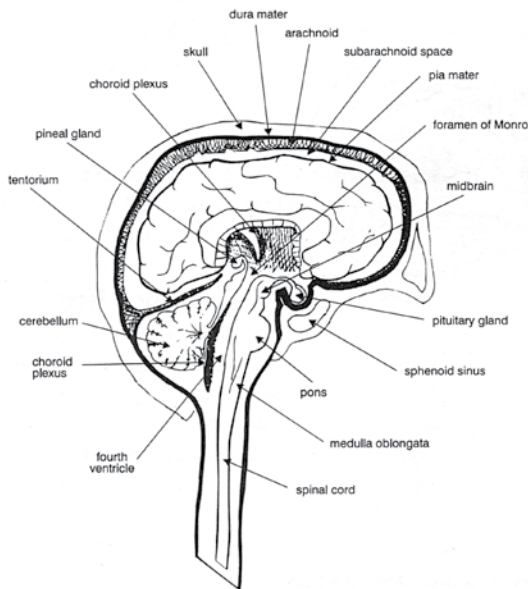
There are two lateral ventricles, one in each cerebral hemisphere. The third ventricle is beneath the corpus callosum and surrounded by the thalamus. The fourth ventricle is an expansion of the central canal of the medulla oblongata.



CROSS SECTION OF THE BRAIN



CENTRAL NERVOUS SYSTEM



SIDE VIEW OF THE BRAIN

AMERICAN BRAIN TUMOR ASSOCIATION INFORMATION, RESOURCES AND SUPPORT

BROCHURES

Educational brochures are available on our website or can be requested in hard copy format for free by calling the ABTA. Most brochures are available in Spanish, with exceptions marked with an asterisk.

GENERAL INFORMATION

About Brain Tumors: A Primer for Patients and Caregivers
Brain Tumor Dictionary*
Brain Tumors Handbook for the Newly Diagnosed*
Caregiver Handbook*

TUMOR TYPES

Ependymoma
Glioblastoma and Anaplastic Astrocytoma
Medulloblastoma
Meningioma
Metastatic Brain Tumors
Oligodendroglioma and Oligoastrocytoma
Pituitary Tumors

TREATMENT

Chemotherapy
Clinical Trials
Conventional Radiation Therapy
Proton Therapy
Stereotactic Radiosurgery*
Steroids
Surgery

AMERICAN BRAIN TUMOR ASSOCIATION INFORMATION, RESOURCES AND SUPPORT

INFORMATION

ABTA WEBSITE | [ABTA.ORG](https://www.abta.org)

Offers more than 200 pages of information, programs, support services and resources, including: brain tumor treatment center and support group locators, caregiver resources, research updates and tumor type and treatment information across all ages and tumor types.

EDUCATION & SUPPORT

- ABTA Educational Meetings & Webinars
In-person and virtual educational meetings led by nationally-recognized medical professionals.
- ABTA Peer-to-Peer Mentor Program
Connect with a trained patient or caregiver mentor to help navigate a brain tumor diagnosis.
- ABTA Connections Community
An online support and discussion community of more than 25,000 members.
- ABTA CareLine
For personalized information and resources, call 800-886-ABTA (2282) or email abta cares@abta.org to connect with a CareLine staff member.

GET INVOLVED

- Join an ABTA fundraising event.
- Donate by visiting [abta.org/donate](https://www.abta.org/donate).

CONTACT THE ABTA

CareLine: 800-886-ABTA (2282)

Email: abta cares@abta.org

Website: [abta.org](https://www.abta.org)

For more information contact:

8550 W. Bryn Mawr Avenue,
Suite 550, Chicago, IL 60631

Website: abta.org

CareLine: 800-886-ABTA (2282)

Email: abta cares@abta.org



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