



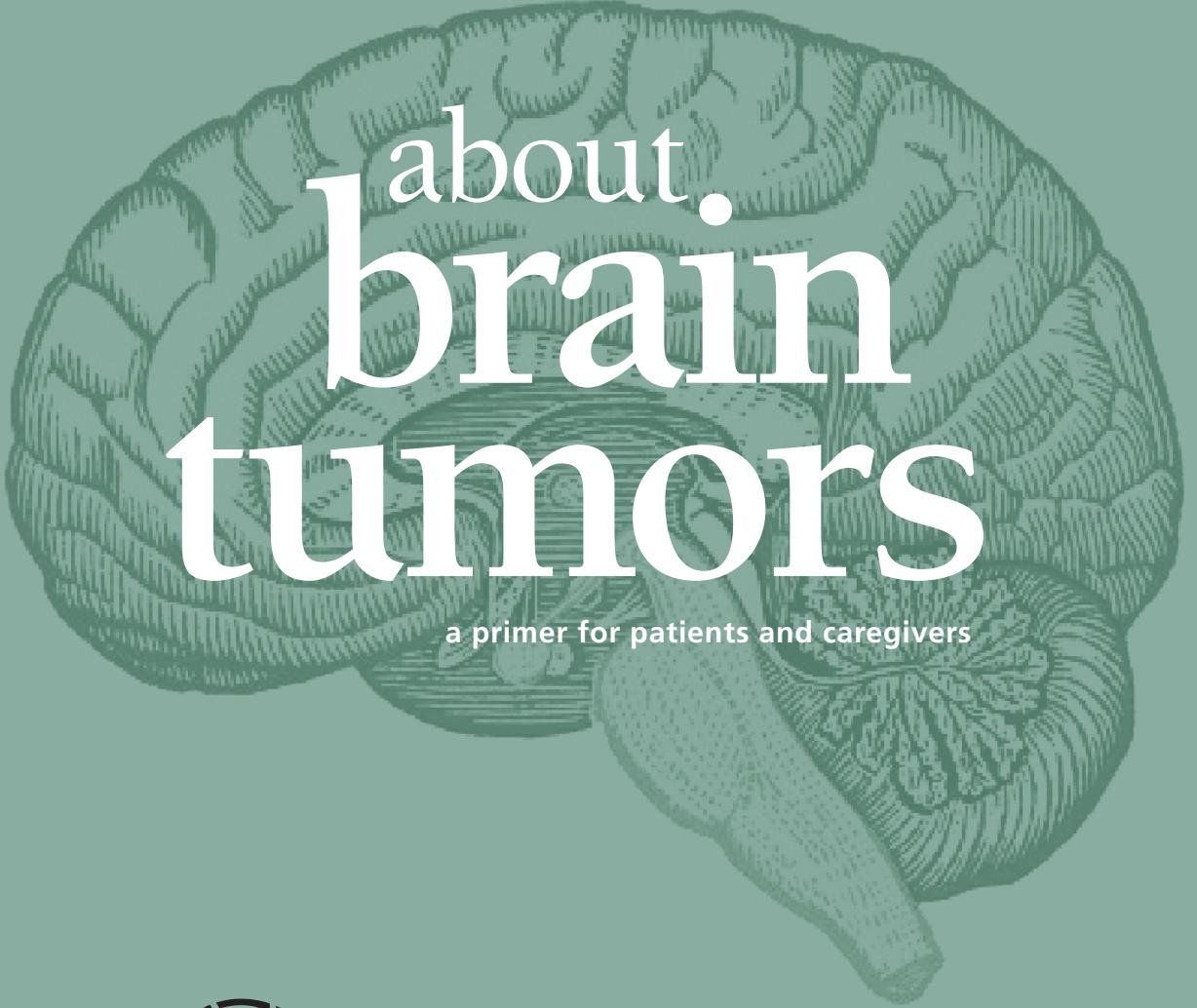
American  
Brain Tumor  
Association®

*Providing and pursuing answers®*



# about brain tumors

a primer for patients and caregivers



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a primer for patients and caregivers



American  
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*Providing and pursuing answers®*

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## 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. For over 40 years, the ABTA has been providing comprehensive resources that support the complex needs of brain tumor patients and caregivers, as well as the critical funding of research in the pursuit of breakthroughs in brain tumor diagnosis, treatment and care.

To learn more about the ABTA, visit [www.abta.org](http://www.abta.org).

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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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# About Brain Tumors

## A Primer for Patients and Caregivers

### 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 health care professionals can provide additional information about tumors, treatment and support resources. We also encourage you to visit our website at [www.abta.org](http://www.abta.org). For more information, please call our CareLine at 800-886-ABTA (2282) or send us an email at [abtacares@abta.org](mailto:abtacares@abta.org).

# Chapter 1: Brain Tumor Basics

Living creatures are made up of cells. The adult body normally forms new cells only when they are needed to replace old or damaged ones. Infants and children create new cells to complete their development in addition to those needed for repair. A tumor develops if normal or abnormal cells multiply when they are not needed.

A brain tumor is a mass of unnecessary cells growing in the brain or central spine canal. There are two basic kinds 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.

When doctors describe brain tumors, they often use the words “benign” or “malignant.” Those descriptions refer to the degree of malignancy or aggressiveness of a brain tumor. It is not always easy to classify a brain tumor as “benign” or “malignant” as many factors other than pathological features contribute to the outcome.

*> 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

A tumor that starts in the brain is a primary brain tumor. Glioblastoma multiforme, astrocytoma, medulloblastoma and ependymoma are examples of primary brain tumors. Primary brain tumors are grouped into benign tumors and malignant tumors.

### Benign brain tumors

A benign brain tumor consists of very slow-growing cells, usually has distinct borders and rarely spreads. When viewed under a microscope, these cells have an almost normal appearance. Surgery alone might be an effective treatment for this type of tumor. A brain tumor composed of benign cells, but located in a vital area, can be considered life-threatening – although the tumor and its cells would not be classified as malignant.

### Malignant brain tumors

A malignant brain tumor is usually rapid-growing, invasive and life-threatening. Malignant brain tumors are sometimes called brain cancer. However, since primary brain tumors rarely spread outside the brain and spinal cord, they do not exactly fit the general definition of cancer.

#### 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*

Malignant brain tumors that are cancerous can spread within the brain and spine. They rarely spread to other parts of the body. They lack distinct borders due to their tendency to send “roots” into nearby normal 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.

### Benign Tumors

- *Slow growing*
- *Distinct borders*
- *Rarely spread*

### Malignant Tumors

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

## 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.”

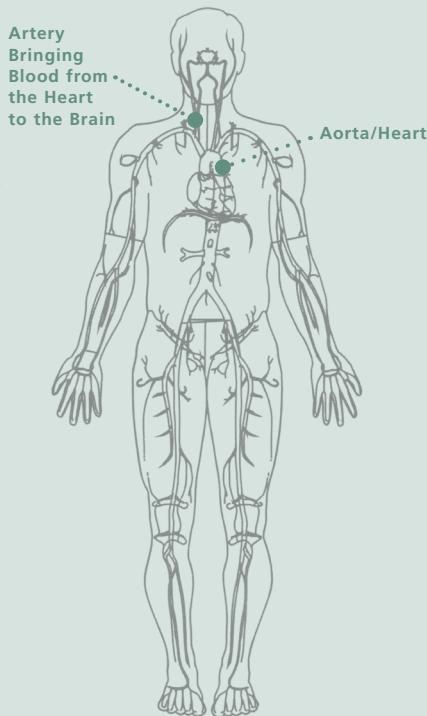
## TUMOR NAMES

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

Tumors are graded to facilitate communication, plan treatment and predict outcomes. The grade of a tumor indicates its degree of malignancy.

Using the WHO grading system, grade I tumors are the least malignant and are usually associated with long-term survival. The 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. Pilocytic astrocytoma, craniopharyngioma and many tumors

**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)*

of neurons – for example, gangliocytoma and ganglioglioma – are grade I tumors.

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.

Grade III tumors are by definition 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.

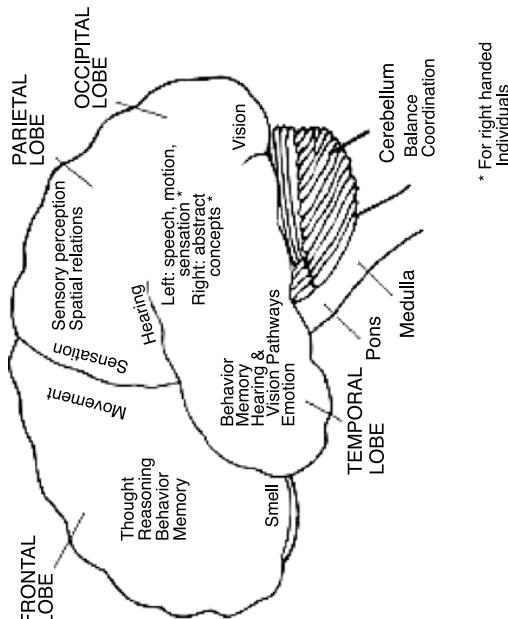
The most malignant tumors are given a grade of IV. They reproduce rapidly, can have a bizarre appearance when viewed under the microscope and easily grow into surrounding normal brain tissue. These tumors form new blood vessels so they can maintain their rapid growth. They also have areas of dead cells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

Tumors often contain several grades of cells. The highest or most malignant grade of cell determines the grade, even if most of the tumor is a lower grade. Some tumors undergo change and a benign 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.

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.

**WHERE IS MY TUMOR?**

- THE NAME OF MY TUMOR IS:**
- Astrocytoma grade I
  - Astrocytoma grade II
  - Astrocytoma grade III, also called Anaplastic Astrocytoma or Malignant Astrocytoma
  - Ependymoma
  - Glioblastoma, also called Glioblastoma Multiforme or Astrocytoma grade IV
  - Medulloblastoma
  - Meningioma
  - Metastatic tumor (primary site: \_\_\_\_\_)
  - Oligodendroglioma
  - Oligoastrocytoma
  - Pituitary Adenoma, also called Pituitary Tumor
  - Other: \_\_\_\_\_

**LOBES OF THE BRAIN****WHEN IS MY NEXT APPOINTMENT? WITH WHOM?**

**MY NURSE'S NAME IS:** \_\_\_\_\_  
Phone: \_\_\_\_\_

**FOR INFORMATION ABOUT BRAIN TUMORS:**

American Brain Tumor Association (ABTA)  
[www.abta.org](http://www.abta.org) 800-886-ABTA (2282)

Cancer Information Service (CIS) of the National Cancer Institute (NCI)  
[www.cancer.gov](http://www.cancer.gov) 800-422-6237

National Institute of Neurological Disorders & Stroke (NINDS)  
[www.ninds.nih.gov](http://www.ninds.nih.gov) 800-352-9424

## WORLD HEALTH ORGANIZATION (WHO) GRADING SYSTEM

### Grade I Tumor

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

### 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 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 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*

## Chapter 2:

# Parts of the Brain

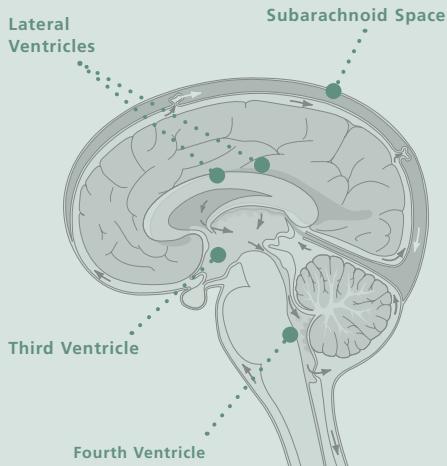
Groups of cells, similar in appearance and with the same function, form tissue. The brain is a soft mass of supportive tissues and nerve cells connected to the spinal cord. Nerves in the brain and spinal cord transmit messages throughout the body. The brain and spinal cord together form the central nervous 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.

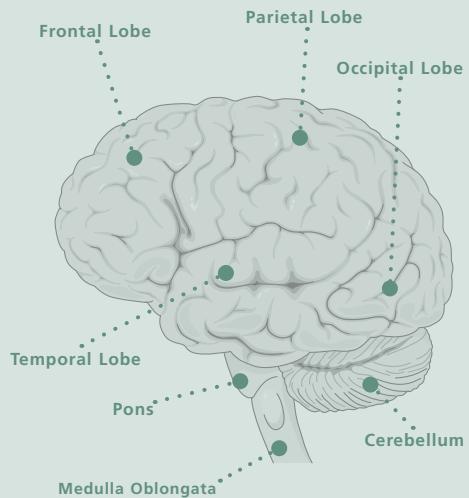
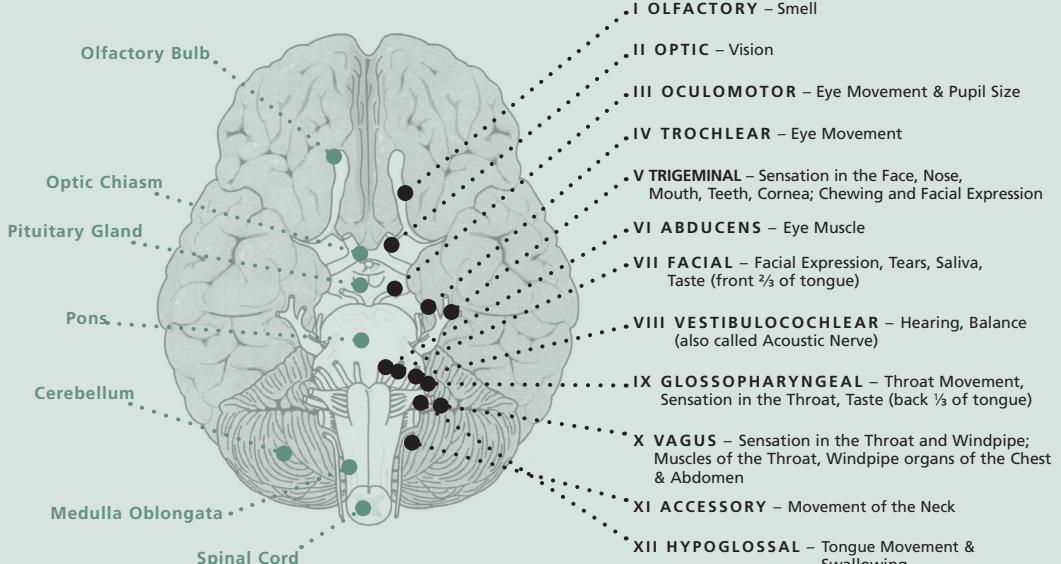
Learning about the normal workings of the brain and spine will help you understand the symptoms of brain tumors, how they are diagnosed and how they are treated.

*Note: For a complete glossary of brain tumor terminology, see Chapter 12 "Brain Tumor Terminology" on page 79.*

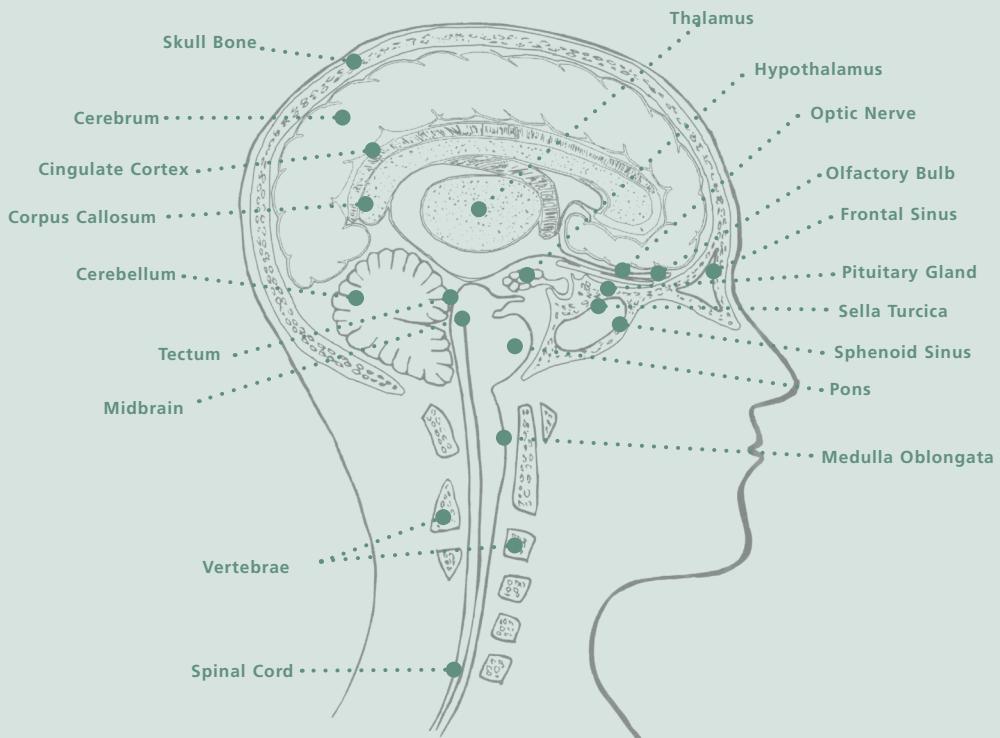
## CSF AND VENTRICLES



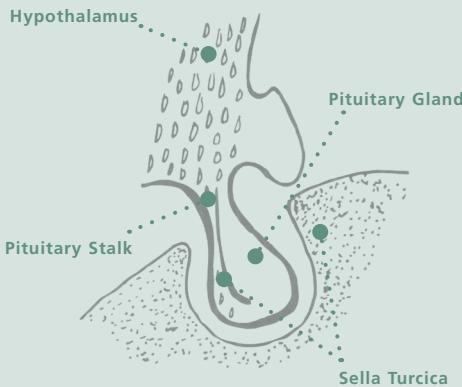
## MAJOR PARTS OF THE BRAIN

CRANIAL NERVES *view from bottom of brain*

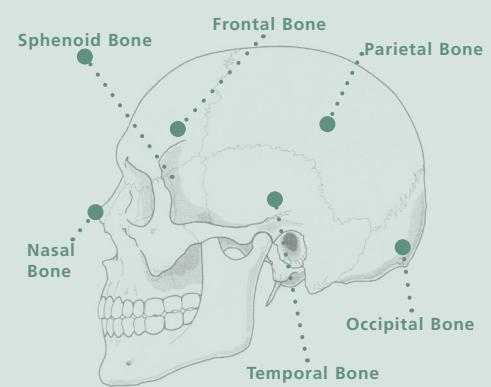
## CROSS SECTION OF THE BRAIN

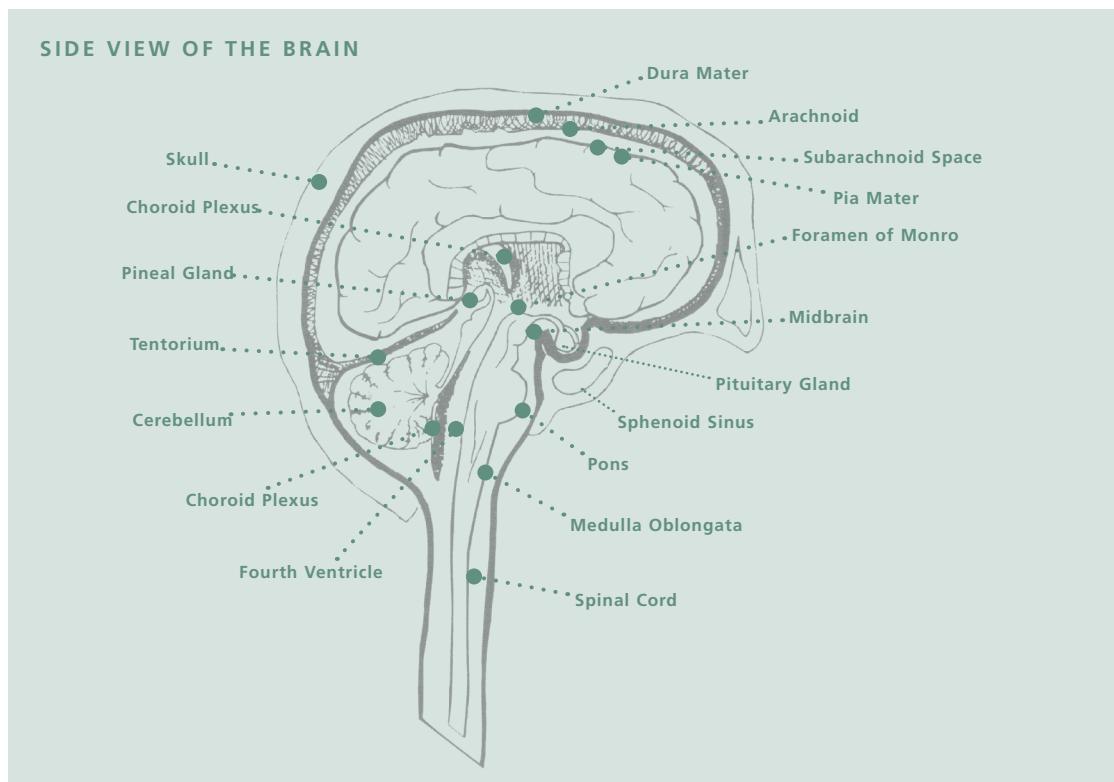
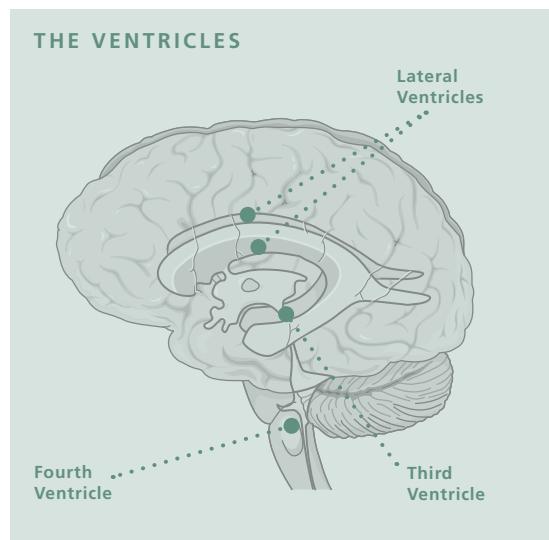
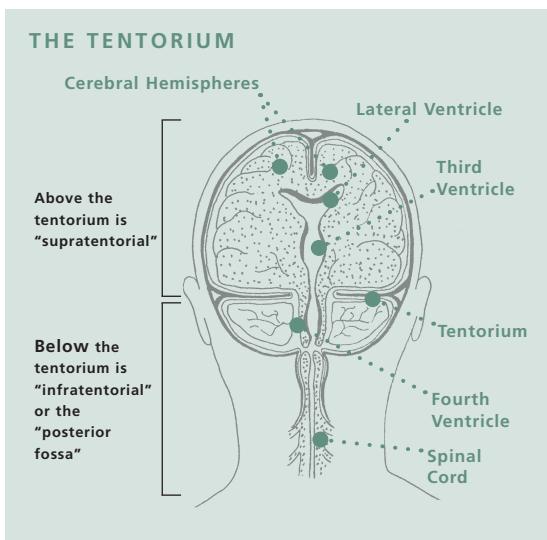


## THE PITUITARY GLAND



## BONES OF THE SKULL





## Chapter 3:

# Types of Brain Tumors

This is an introduction to the more common brain tumors, their typical symptoms and locations and how they might be treated. Please remember that your tumor is unique and might not conform to the “average” characteristics described.

The tumor names and their organization in this chapter are based on the World Health Organization (WHO) brain tumor classification system.

For more information about specific tumors and treatments visit [www.abta.org](http://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 include pilocytic astrocytomas, which are usually localized (limited in growth) tumors, that are often cured through surgical removal. Grade II to IV tumors have increasing degrees of malignancy and although surgery is beneficial, it is not curative for 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 and anaplastic astrocytoma.

### Pilocytic Astrocytoma

#### *Also called juvenile pilocytic astrocytoma*

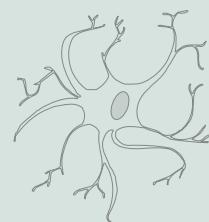
These grade I astrocytomas are usually well-defined, non-infiltrating tumors – meaning they tend to stay in the area in which they started and do not spread into surrounding tissue. They generally form cysts or may be enclosed within a cyst. Although these are usually slow-growing tumors, they can become 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 (an “optic glioma”), the optic chiasm near the hypothalamus, thalamus, basal ganglia, cerebral hemispheres and the cerebellum (i.e., a cerebellar astrocytoma).

This tumor is the “most benign” 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

### ASTROCYTE

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



cannot be completely removed. Or, the residual 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 benign pilocytic astrocytoma.

### 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 cells and cell grades, but brain tumors are graded by the highest grade (most abnormal) cell seen in the tumor. These tumors tend to have tentacle-like projections that grow into surrounding tissue, making them difficult to completely remove during surgery.

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 the surrounding tissue, these tumors cannot be totally removed during surgery. Partial removal can help decrease symptoms; the tissue obtained during that surgery confirms 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 margin 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, implanted radiation and proton therapy – which may be recommended to you. Your radiation oncologist will decide which form of radiation therapy is best for your particular tumor.

Chemotherapy, most often with the drug temozolomide, may be recommended immediately after radiation or when the tumor recurs. Some treatment plans may still use the drugs BCNU, CCNU, procarbazine or cisplatin. In addition, there are also many new drugs being tested in clinical studies (trials). Some physicians may choose not to use chemotherapy for the initial tumor, “reserving” it for re-growth if necessary. 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 “Glioblastoma” on page 21.

## BRAIN STEM GLIOMA

Brain stem gliomas arise in or on the brain stem – the area containing all of the converging 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 and 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 determination of grade impossible. In these situations, the diagnosis can usually be based on the MRI scan features.

Most of these tumors are classified by their location:

Cancer is a disease defined by:

- Upper brain stem (midbrain or tectum)
- Middle brain stem (pons)
- Lower brain stem (cervico-medullary)

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 precluding the tumor’s surgical removal. A few of these tumors are localized and may be reachable for resection. These tumors tend to be very slow growing, not located in the pons and exophytic.

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) causing headache, nausea, vomiting and gait unsteadiness.

Treatment of a brain stem glioma is dictated by the tumor location, grade and symptoms. Surgery may be warranted if a tumor appears circumscribed (contained) or exophytic. The goals of surgery are to determine the grade and type of tumor and, sometimes, removal of 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 may be considered.

Radiation therapy with hyperfractionation (with smaller dose per treatment and many more doses) 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. Clinical trials using various forms, doses and schedules of radiation therapy for newly diagnosed tumors and chemotherapy for recurrent tumors, are also available (see “Clinical Trials” on page 23).

### **CRANIOPHARYNGIOMA**

This is a benign 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. Malignancy and metastasis are unknown.

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

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 – or a radiation source may be implanted into the tumor cavity, such as radioactive phosphorous. 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.

## EPENDYOMA

Recent studies show that 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 1–2% of all primary tumors and 6–7% 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), *myxopapillary ependymomas* and *ependymomas* (grade II) and *anaplastic ependymomas* (grade III). The grade is based on how much the cells look like normal ependymal cells, although various grading systems exist. The cells of a grade I tumor look somewhat unusual, whereas grade IV tumor cells look definitely abnormal.

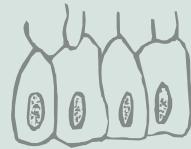
**Subependymomas** usually occur near a ventricle. **Myxopapillary ependymomas** tend to occur in the lower part of the spinal column. Both of these ependymoma types are slow growing and are considered to be low-grade or grade I tumors.

**Ependymomas** are the conventional type, lowgrade (grade 1 and II) tumors. These tumors 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 *tanyctic ependymoma*. There are several other patterns as well, but regardless of appearance, these are all considered grade II tumors.

**Anaplastic ependymomas** are high-grade tumors (grade III) and tend to be faster growing than the low-grade tumors. These are most commonly

### EPENDYOMA

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



found in the brain in adults and specifically in the posterior fossa in children. They are rarely found in 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 following surgery if all visible tumor wasn't removed and in some cases even after complete resection. If the tumor is localized, 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 very young children.

## GERM CELL TUMORS

These uncommon tumors represent 1–3% of childhood brain tumors 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 with gadolinium enhancement 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 detect recurrence.

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

## GLIOBLASTOMA

*Also called “astrocytoma, grade IV” and “GBM”* “Grade IV astrocytoma,” “glioblastoma” and “GBM” are all names for the same tumor. This tumor represents about 16% of all primary brain tumors and about 60–75% of all astrocytomas. They increase in frequency with age and affect more men than women. Only three percent of childhood brain tumors are glioblastomas.

Glioblastomas are generally found in the cerebral hemispheres of the brain, but can be found anywhere in the brain or spinal cord. Because glioblastomas can grow rapidly, the most common symptoms are usually due to increased pressure in the brain and can include headache, nausea, vomiting and drowsiness. Depending on the location of the tumor, patients can develop a variety of other symptoms such as weakness or sensory impairment on one side of the body, seizures, memory or language impairment and visual changes.

Glioblastomas commonly contain a mix of cell types. It is not unusual for the tumor to contain cystic material, calcium deposits, blood vessels or a mixed grade of cells. The diagnosis of a glioblastoma is based on several features when the tissue is examined: the cells are highly malignant, there are abnormal and numerous blood vessels and a high percent of tumor cells are reproducing at any given time. Necrotic (dead) cells may also be seen, especially toward the center of the tumor. The growing blood vessels may be seen throughout the tumor, but are generally present in highest number near the edges of the tumor. These blood vessels bring nutrients to the tumor, assisting in its growth. Since these tumor cells arise from normal brain, they easily intermingle with and invade normal brain tissue. However, glioblastoma rarely spreads elsewhere in the body.

In recent years, advanced biotechnology has allowed glioblastomas to be sub-divided into two groups: primary and secondary glioblastoma. Primary or de novo, glioblastoma arise quickly and tend to make their presence known abruptly. These are the most common, very aggressive form of glioblastoma. Secondary glioblastoma may have a longer, somewhat slower growth history but are still very aggressive tumors. These glioblastoma may begin as lower grade tumor and then transform into higher grade. They tend to be found in people ages 45 and younger and represent about 10% of the glioblastomas. Scientists are now developing tests that may help better identify these two sub-categories of glioblastoma. That information may also soon lead to other sub-groupings of glioblastoma and therapies specific to those biological differences between tumors. However, it does not appear that there are any differences in prognosis for either of these types of glioblastoma.

Lack of exactly the same cells from end to end of the tumor makes a glioblastoma one of the most difficult brain tumors to treat. While one cell type may be responsive to treatment, other types may

be resistant. For this reason, the treatment plan for glioblastoma will combine several approaches.

The first step in treating a glioblastoma is surgery to make a diagnosis, relieve pressure and safely remove as much tumor as possible. Because these tumor cells have octopus-like tentacles, there are no clear edges to glioblastomas. This feature makes them very difficult to remove completely. If the tumor is located near important structures such as the language center or motor area, the ability to remove most of the tumor may be further limited.

Radiation therapy, accompanied by chemotherapy, almost always follows surgery or biopsy. Radiation therapy affects mostly replicating cells and therefore causes more damage to tumor cells than to normal brain cells (most cells in the brain are not actively dividing). The most common type of radiation is called fractionated external beam radiation, meaning that the radiation is given in several treatments over a few weeks. (This is also called standard radiation or conventional radiation.) It is given to the tumor and a margin around it, but not to the whole brain. Another type of radiation sometimes used for glioblastomas is conformal or intensity modulated radiation therapy (IMRT). Other types of radiation may be used on an experimental basis but are not considered “standard” therapies (for example brachytherapy, which consists of either implanted radioactive seeds or catheters with temporary radioactive sources in the tumor or monoclonal antibodies tagged with radioactive particles). Because of its very focused beams and the need to radiate some amount of tissue around the central mass of a glioblastoma, stereotactic radiosurgery is generally not used for this tumor. The exception may be in treating a tumor with a very specific, localized area of growth or regrowth. In that situation, radiosurgery may be used as a “boost” to that very confined area; however this also is a strategy that is not widely used.

The most commonly used chemotherapy drug in adults is currently temozolomide; however, other drugs are also being tested. Many of the studies combine temozolomide with other drugs which have different biological actions, such as those affecting blood vessel growth or drugs which interfere with proteins created by the tumor. Some neurosurgeons use biodegradable wafers which contain the chemotherapy drug BCNU. The wafers are placed in the cavity created during tumor removal. Other new delivery systems which place drug directly into the tumor area are under investigation as well. Chemotherapy might also be used to delay radiation in young children.

Because glioblastoma cells tend to move into nearby tissue, total removal of these tumors is not possible. Tumor regrowth can be treated with additional surgery, another form of focused radiation, a different chemotherapy drug or combination of drugs or any number of new approaches to these tumors.

An area of active research interest is the development of drugs that target specific biological abnormalities found in the tumor cells. Many of these drugs interfere with or block signaling pathways within the tumor – the message patterns tumors and their byproducts (such as proteins or enzymes) create. The ability to identify these biologic differences and to create drugs that target these differences, are called “personalized” or “individualized” medicine. While this is an exciting area of science, development and testing of these drugs are in the earliest stages.

Immunotherapy – the use of vaccines or immunizations – is another area of research interest. These therapies use the body’s own immune system to fight a tumor. There are several research studies focusing on this area of treatment and many of these studies are open to those with a glioblastoma. Some of these treatments use tumor cells, removed at the time of surgery, which are treated in a laboratory then re-injected

as a “vaccine” back into the patient. The goal of these treatments is to trigger the body’s immune system into mounting a response to the tumor. Some vaccines combine the treated tumor cells with a drug or other substance. Immunotoxins, such as diphtheria or pseudomonas, link a toxin to a radioactive antibody and carry it to the tumor cells. Monoclonal antibodies combine a radioactive substance with a substance that will trigger an immune response. These new therapies are offered in organized research studies called clinical trials.

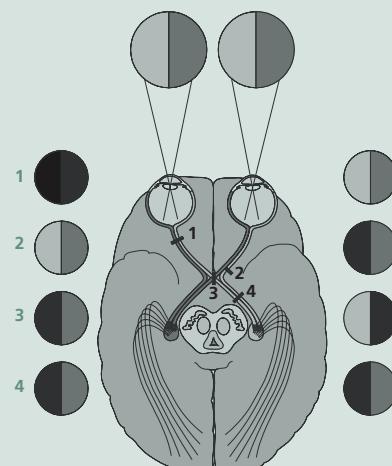
### Clinical Trials

A clinical trial 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 be unavailable to them.

Some clinical trials are targeted to certain patients based on the genomics of their specific tumor. Genomic testing is available for patients commercially and at some of the larger brain tumor institutions.

The ABTA can assist patients who want to better understand the risks and benefits of clinical trials, patient rights and protections in a trial and more. In addition, ABTA offers TrialConnect®, a free, confidential service that links brain tumor patients with appropriate clinical trials based on the patient’s tumor type and treatment history. For more information on clinical trials, visit [www.abta.org](http://www.abta.org), call the ABTA CareLine at 800-886-ABTA (2282) or send an email to [abtacares@abta.org](mailto:abtacares@abta.org). For more information on TrialConnect®, visit [www.abtatrialconnect.org](http://www.abtatrialconnect.org), call 1-877-769-4833 or click on the TrialConnect® link on the ABTA website.

### VISUAL CHANGES WITH TUMORS ALONG THE OPTIC PATHWAY



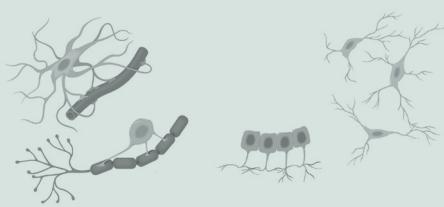
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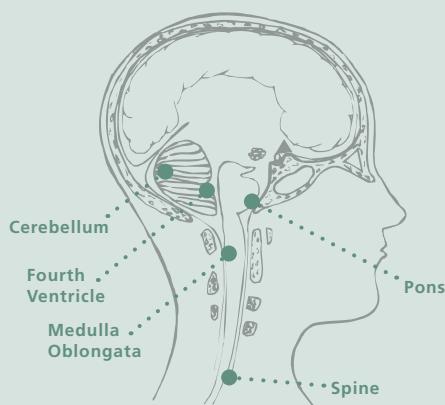
### GLIOMA

This is a general term for any tumor that arises from the supportive, or gluey, tissue of the brain. This tissue, called glia, 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. An astrocyte (star-shaped cell) will give rise to astrocytomas (including glioblastomas), an oligodendrocyte (cell with short arms forming the insulation of neurons) will give rise to oligodendrogiomas and lastly, tumors called ependymomas arise from ependymal cells (i.e., the cells that form the lining of the fluid cavities in the brain). Occasionally, tumors will display a mixture of these different cells and are called mixed gliomas (see “Mixed Gliomas” on page 26).

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.

### GLIAL CELLS



**LOCATION OF THE CEREBELLUM****MEDULLOBLASTOMA**

Medulloblastomas represent about 13% of the brain tumors in children under the age of 14. In addition, medulloblastomas represent about 3% of the brain tumors in adults. Medulloblastomas are always located in the cerebellum.

Medulloblastoma is a fast-growing, high-grade tumor which 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; gait unbalance and poor coordination of the limbs; and unusual eye movements.

Treatment consists of surgical removal of as much tumor as possible, radiation and chemotherapy. Testing will also be done to check for possible tumor spread, including an MRI of the spine and a cerebrospinal fluid analysis. For older children, adults without evidence of the tumor spreading

and those for whom most of the tumor has been removed, radiation to the tumor area followed by a lower dose of radiation to the entire brain and spinal cord follows surgery. Very young children are often treated with chemotherapy instead of radiation to defer its use until they are older.

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.

New therapies and new treatment plans are developed in organized programs called clinical trials (see “Clinical Trials” on page 23).

**METASTATIC BRAIN TUMORS**

A metastatic or secondary, brain tumor is formed by cancer cells from a primary cancer elsewhere in the body which spread to the brain. In most situations, the primary cancer is diagnosed before it spreads to the brain, but in some circumstances the brain tumors are found the same time or before the primary cancer is found. Cancers that frequently spread to the brain include:

- Lung cancer
- Breast cancer
- Melanoma (malignant skin cancer)
- Kidney cancer
- Colon cancer

**MENINGIOMA**

These tumors arise from the “arachnoid mater” – one of the layers of the meninges (the lining of the brain). Meningiomas represent about 38% of all primary brain tumors and occur most frequently in middle-aged women. The majority

of meningiomas are benign, grade I, slow-growing tumors which are localized and non-infiltrating. Meningiomas are most often located between the cerebral hemispheres (“parasagittal meningiomas”); within the meninges, the protective tissues that cover the spinal cord and brain (“convexity meningiomas”); at the base of the skull; and 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.

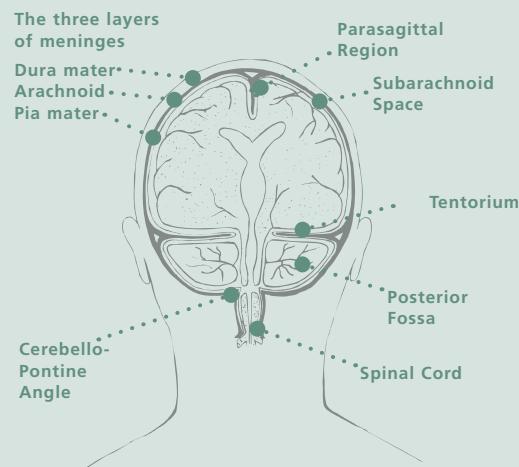
A variety of symptoms are possible, depending on the tumor’s location. The most common indications are headache, weakness on one side, seizures, personality and behavioral changes and confusion. Neuro-imaging (scanning) with a CT or MRI is used to evaluate the location of the tumor. Calcifications may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

The benign meningioma (grade I) is slow-growing with distinct borders. Because it grows slowly, it

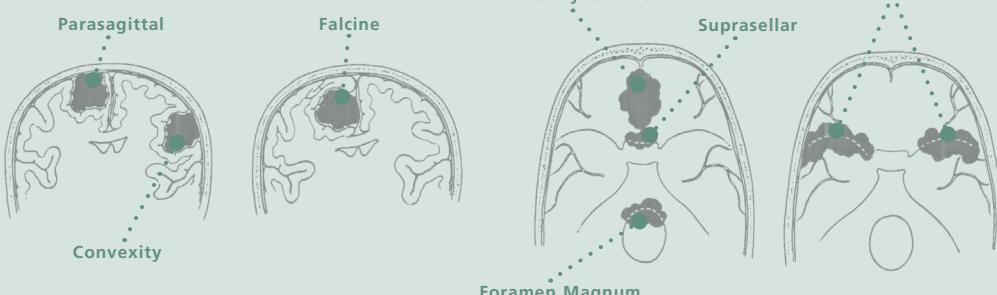
can grow quite large before symptoms become noticeable. Symptoms are caused by compression rather than by the tumor growing into brain tissue.

If the tumor is accessible, the standard treatment is surgery to remove the tumor, the portion of the dura mater (the outermost layer of the meninges) to which it is attached 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 preoperatively and in some cases the blood vessels are embolized (purposefully blocked) to facilitate the removal

### MENINGES



### 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.

of the tumor. Radiation therapy or radiosurgery might be of value if the tumor is not entirely removed. For some patients, surgery may not be recommended. For those with no symptoms (when they have been diagnosed coincidentally), those with minor symptoms of long duration and those for whom surgery would be risky, long-term close observation with scans may be advised. An alternative includes focused radiation, also called “stereotactic radiosurgery.”

The atypical meningioma (grade II) has a middle range of behavior. These tumors are not clearly 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. Drugs that target abnormal signaling pathways within the tumor are also being evaluated. Hormone therapy does not appear effective.

### MIXED GLIOMA

Mixed gliomas commonly contain a high proportion of more than one type of cell. Most often these tumors contain both astrocytes and oligodendrocytes – these tumors are generally called mixed gliomas or oligoastrocytoma. Occasionally, ependymal cells are also found. The behavior of a mixed glioma tumor tends to be

based on the grade of the tumor. It is less clear whether the tumor behavior is closer to that of the most abundant cell type.

Standard treatment for a mixed glioma is similar to that for an astrocytoma and oligodendrogloma 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 “short arms” or a fried-egg shape as opposed to astrocytomas, which have “long arms” or a star-like shape.

Oligodendroglomas can be low-grade (grade II) or high-grade (grade III also called anaplastic). Sometimes oligodendroglomas may be mixed with other cell types. These tumors may also be graded using an “A to D” system which is based on microscopic features such as the appearance of the cell nucleus, the number of blood vessels and presence or absence of dead tissue called necrosis. The grade denotes the speed with which the tumor cells reproduce and the aggressiveness of the tumor.

Oligodendroglomas occur most frequently in young and middle-aged adults, but can also be found in children. The most common location is the cerebral hemisphere, with about half of those tumors being found in the frontal lobe. Seizure is

#### OLIGODENDROCYTE

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



the most common initial symptom, particularly in low-grade tumors.

Standard treatment for accessible tumors is surgical removal of as much tumor as possible. Biopsy alone may be performed for inaccessible tumors – those that cannot be surgically removed. The tumor sample removed during a biopsy is used to confirm the diagnosis and the grade of tumor.

For low-grade oligodendrogloma 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 appears to be indicated although the best timing – immediately or at tumor progression – is being determined in clinical trials. Recurrent low-grade oligodendroglomas can be treated with surgery, radiation therapy (if not given initially) or chemotherapy.

For anaplastic oligodendrogloma, a combination of radiation therapy and chemotherapy such as PCV (procarbazine, CCNU and vincristine) or temozolomide is indicated. Recurrent anaplastic oligodendrogloma may be treated with surgery and/or chemotherapy. Genetic analyses of oligodendrogloma 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. Clinical trials are available for newly diagnosed and recurrent, low-grade or high-grade oligodendroglomas. Many of these trials take into account the genetic features of the tumor, thereby highlighting the importance of obtaining tumor tissue for analysis (see “Clinical Trials” on page 23).

## 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 oligodendroglomas) 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 radioactive implants or 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. For more information on clinical trials, see “Clinical Trials” on page 23.

## ADDITIONAL TUMOR TYPES

Other brain tumor types include:

- Acoustic Neuroma
- Atypical Teroid Rhabdoid Tumor (ATRT)
- Chondroma, Chondrosarcoma, Chordoma
- Choroid plexus tumors
- Cysts
- Dysmbyoplastic neuroepithelial tumor (DNT)
- Gangliocytoma
- Ganglioglioma
- Germinoma
- Glioma, optic
- Gliomatosis cerebri
- Glomus jugulare
- Hemangioblastoma
- Hemangipericytoma
- Lipoma
- Neuroblastoma cerebral
- Neurocytoma central
- Neurofibromatosis
- Pineal

- Pituitary
- PNET
- Pseudotumor cerebri
- Schwannoma
- Skull base
- Spinal cord
- Teratoma
- Tuberous sclerosis
- Vestibular schwannoma

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

## 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 would explain why people develop brain tumors and what these people have in common with each other. These observations of “commonality” can provide important clues as to the links between individuals. Once one of these findings has been replicated by other scientists or additional studies – a process called validation – then this finding would be considered a convincing cause or risk factor for that disease.

## INTRODUCTION

Causes and risk factors can be *environmental*, such as being exposed to poisonous substances in the home or at work; eating or not eating certain foods; or whether or not we exercise, smoke cigarettes or drink alcohol. They can also be *genetic*, such as being born with a gene mutation or susceptibility that one inherits from parents. These genetic mutations/susceptibilities may also accumulate over time, as one grows older.

Unfortunately, no risk factor accounting for the majority of brain tumors has been identified, 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, second hand smoke, agricultural chemicals and industrial formaldehyde
- 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 antihistamines
- 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 development of brain tumors.

Of particular interest over the last decade has been the potential association between cell phone use and risk of developing a brain tumor. 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 (>10 years) cell phone use with further conflicting results. In general, the conclusions from most of these studies are (1) there is no consistent association between cell phone use and risk of developing a brain tumor (benign or malignant) and (2) there is a very slight increased risk of a brain tumor associated with using a cell phone for 10 years or more. Further studies, in both the laboratory and in humans with longer follow up, are needed to fully understand this exposure and any potential relationship with brain tumor development.

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 then moved to the brain) or primary brain tumors (those that began in the brain and tend to stay there). If the brain tumors are primary tumors, scientists will want to know the specific type(s) of primary brain tumors. The clusters of most concern are those involving the same type of primary brain tumor, since these tumors may share similar biologic origins. Metastatic brain tumors, such as breast cancer, lung cancer or colon cancer that has 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% of all cancer is actually inherited from one generation to another in a family (also called hereditary). There are a few rare, inherited genetic syndromes that involve brain tumors. Hence, there are very few families where multiple people in that family have a brain tumor. In those syndromes, a mutation in a specific gene is 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).



- “*Genetic*” means anything referring to our genes.
- “*Inherited*” or “*hereditary*” means a disease or condition passed from one generation to another within a family.

The vast majority of genetic risk factors, however, are not inherited at birth but actually accumulate over time as we age (also called somatic or acquired). While most of our genes go about their jobs as expected, a small number may become inactive or begin 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:

- *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. EGFR is a growth factor that has been well studied in brain tumors and has been shown to be 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.

With the publication of the Human Genome 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. Two recent GWA studies of glioma found some results in common, but they also found some differing results. The scientists involved in these studies believe the differences in their results may be due to the differences in the people who were part of their studies. This research shows that common genetic differences amongst the 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 benign brain tumors or pediatric brain tumors.

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

#### The Cancer Genome Atlas (TCGA) Project

Studies of any specific gene are complicated by the fact that there are many potential genes in the human genome to consider. 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 National Human Genome Research Institute (NHGRI), has a goal of completely cataloging all of the somatic genetic changes in more than 20 different cancers, then making these data publically available in order to improve the ability to diagnose, treat and prevent cancer. TCGA started as a pilot project in 2006 prioritizing glioblastoma (GBM), ovarian and lung cancers as the first cancers to study. The first GBM paper published under this project showed three biological pathways involved with GBM. Since that publication, other scientists have described additional key genetic changes associated with malignant brain tumors. Some of these reports include important comparisons with low grade gliomas and other glioma subtypes. TCGA is now expanding its efforts to include other types and grades of gliomas.

#### Chromosome Changes

Another area of scientific study is the ability tumors have to lose or gain pieces of *chromosomes*. Each normal cell in any human body has 23 pairs of chromosomes. The most common chromosomal changes in brain tumors occur on chromosomes 1, 10, 13, 17, 19 and 22.

Changes on chromosomes 1 and 19 are most frequently found in oligodendrogiomas. 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.

### 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. The ABTA's resources are available to help you locate a genetic counselor.*
- *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 in 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 the 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 "deja 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. Double vision, 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 vision 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.

**Brain Stem Tumors** often cause vomiting and a clumsy gait. 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. The brain stem also controls vital life functions such as breathing and heartbeat.

**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, 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 in 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 cancer 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 health care 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,” on page 63.

## 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. Additionally, a listing of wig and head covering resources is available through the ABTA. 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 health care 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 travel 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](http://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 – 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 probably the most routinely used imaging technique for diagnosis and follow-up of many tissue

abnormalities. 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 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

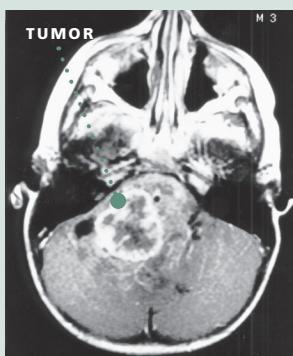
signals, the computer can distinguish between healthy and diseased tissue.

Patients with some cardiac monitors, pacemakers or some types of surgical clips cannot 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).

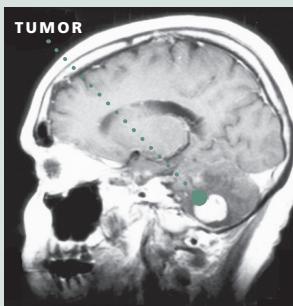
An MRI offers images with excellent anatomical detail that provides clarity of the small structures in the brain, but the images often lack quantitative or finely measurable, information. Because of this lack of very fine measurable detail, it can take a while before the effectiveness of drug therapies can be imaged. Researchers are working toward new scanning techniques that will more rapidly image treatment effects.

#### MRI SCAN



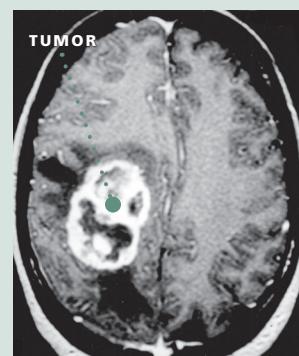
This MRI scan shows an ependymoma.

Scan courtesy of Dr. Regina Jakacki



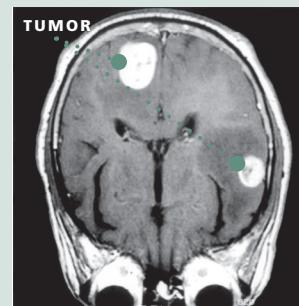
MR scan of a single breast metastasis in the cerebellum.

Scan courtesy of Dr. Deborah Heros



MR scan of a glioblastoma multiforme.

Scan courtesy of Dr. Jeffrey Bruce



MR scan of multiple brain metastases.

Scan courtesy of Dr. Raymond Sawaya

### Other CT or MRI Based Scans

Computer technology advances have made possible the development of new methods for using existing scanning equipment. These new methods provide advanced tools for diagnosis.

### 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 (such as drugs) 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.

### 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.

### MRS (Magnetic Resonance Spectroscopy)

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 and in differentiating between tumor recurrence and radiation necrosis. This technique may also be valuable in suggesting the degree of malignancy. MRS and PET are complementary tools for metabolic imaging.

**PET (Positron Emission Tomography)**

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 (as we think, our brain uses sugar as fuel) causing considerable background color in the PET images. Other radioactive substances 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, measurements of brain or brain tumor activity (determined by concentrations of the radioactive substance) are fed into a computer, which produces a color-coded moving picture of the brain as it accumulates the radioactive sugar or drug. The use of PET had been somewhat limited because the equipment is expensive and requires radioactive materials (drugs) 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 the community.

**SPECT (Single Photon Emission Computerized Tomography)**

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

A SPECT scan is similar to PET. Radioactive tagged materials taken up by the brain are used. A special camera measures the rate of emission of the material as it moves through the brain. Images are generated from that information. After MRI or CT, this test might be helpful in distinguishing between low-grade and high-grade tumors or between recurrent tumor and necrosis.

**MEG (Magnetoencephalography)**

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 help scientists identify the way the parts of the brain interact with each other, how the brain processes information and the pathways followed by information as it enters the brain. This may also help us understand why certain brain tumors, based on their location, cause specific functional problems.

The device 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 very limited number of facilities. As government support for the development of this technique increases, community access may also increase.

*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.*

## X-RAYS

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

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. Only examination of a sample of tumor tissue under a microscope provides an exact diagnosis.

## LABORATORY TESTS

### Biomarker Research

Recent advances in 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 minuscule bits of material are being explored for their potential use in diagnosis, treatment and 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 aggressiveness of the 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 the brain's bulging through an opening in a membrane, muscle or bone (herniation).

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 meningeal tumor. After surgery, 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.

The CSF may also be examined for the presence of known tumor markers, in addition to tumor cells and substances that indicate the presence of a tumor. Scientists are working toward identifying and characterizing the biomarkers for brain tumors. Biomarkers for germ cell tumors are well-known. They include:

- AFP (alpha-fetoprotein)
- HCG (human chorionic gonadotropin)
- PLAP (placental alkaline phosphatase)

CEA (carcinoembryonic antigen) is a marker for a tumor of the arachnoid and/or pia mater membranes of the meninges (a leptomeningeal tumor). These are usually metastatic tumors.

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

### Myelogram

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 many 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.

#### **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 study and analysis. Only then is a tissue 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

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

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.

## GENOMICS

Doctors are now using genomic changes in the tumor to identify sensitivity to treatment and prognosis. This information is being more commonly collected from tumor samples at biopsy or tumor removal. Some clinical trials are targeted to certain patients based on the genomics of their specific tumor. Genomic testing is available for patients commercially and at some of the larger brain tumor institutions.

## 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:

- Be aware that classification of brain tumors by the pathologist is a subjective procedure that is not always straightforward. Different pathologists might disagree about the classification and grade, of the same tumor.
- 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 new differences and new similarities in tumors. Sometimes this means re-naming or re-grouping tumors.

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 1–3 months after surgery and/or the completion of radiation therapy. This time gives the brain a chance to begin healing from the effects of surgery or radiation. Although it can be difficult to wait, scans done during this time would most likely show the swelling that can occur in this time period 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 3 months, 6 months or perhaps yearly. The follow-up should 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. Other tests help evaluate the effectiveness of medications, such as antiepileptic (anti-seizure) drugs.

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

## PROGNOSIS

Prognosis means prediction. It is an educated guess about the future course of a disease in a specific individual.

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 benign 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 lots of 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 number 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 off questions from the list and writing down responses.

If it's 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 doctors' visits, 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 don't understand. Or perhaps you would like guidance about resuming your routine activities.

We encourage you to take these questions to your health care 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're participating in your health care. By gathering information, you'll 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 illness. 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 health care 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.

Not everyone wants or needs a second opinion. Yet, 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.

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's 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'll 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 and consultation reports, as well as 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 store 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're the patient or a family member, it's 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. They 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 health care team.

## ABOUT INSURANCE

After your first visit, you'll need to verify your health care insurance coverage. The answers to

most of your insurance questions can be found in the insurance policy itself or the policy manual. If you don't have a copy, now is the time to obtain one.

For employer-provided health insurance, contact your employer's Human Resources office 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?"

## Chapter 7:

# Seizures

Seizures are common symptoms of a brain tumor. Between 25 and 40% 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, and fever can all cause seizures. Or, the cause may 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, menses, increased stress, alcohol, new medication or changed medication doses all can be triggers. Keeping a diary or journal of activities and feelings that occur prior to each seizure can help identify personal triggers.

What does someone having a seizure look like? This is a normal question. 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." Still, for others, a seizure can take the form of 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 him 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 health care 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 with whom you spend time, can help prepare them as well. Remember that most seizures end naturally.

Your role becomes remaining calm and protecting the person from environmental harm at a time they cannot protect themselves. 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
- The seizure lasts more than 5 minutes
- A second seizure immediately follows
- The seizure occurs in water

## 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, accompanied by or followed by psychic symptoms. 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 are 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. Dieticians 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 ANTIEPILEPTIC 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. A double dosage should not be taken if a dose is missed. Instead, the regular schedule should be resumed and the doctor notified. *If you stop taking your medicine abruptly, seizure activity will increase. Call your doctor for assistance if you miss more than one dose, notice an increase in your seizures or develop a rash.*

### Check Levels if Indicated

Some medications require frequent blood tests in order to check the drug levels in the body. Ask the doctor if the medication should be monitored in this way. If yes, 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 the antiepileptic medications will be needed. The decision is based on the seizure history,

how often the 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 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 doesn't control the 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

For your own protection, 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

Your gums may be inflamed, red, swollen, tender or bleeding. Good oral hygiene, with regular brushing and flossing, is key in managing this side effect that is influenced by bacteria levels in the mouth. 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

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.

**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 medications is always a medical emergency.*

**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?
- 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.

7

**Stress**

You may be able to reduce stress through exercise, meditation, yoga, guided imagery, deep breathing and/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 neurological disorders can have on a patient and their family.

**Relationships**

Yes, seizures can be stressful to 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. Or, consider reaching out to a neuropsychologist – a professional trained in the workings of the brain and the psychological impact neurological disorders can have on a patient and their family.

**Driving**

Laws prohibiting people with seizures from driving are designed to protect both you and other people from injury. Talk with a licensed health care 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.

For more information about seizures, including "Seizure First Aid Tips" clings for the home or office and tips for living with seizures, visit [www.abta.org](http://www.abta.org), call 800-886-ABTA (2282) or send an email to [abtacares@abta.org](mailto:abtacares@abta.org).

## Chapter 8:

# Pediatric Brain Tumors

Children are not smaller versions of adults. Their bodies and brains are still developing. Their needs are different. 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% 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 full listing of *pediatric* tumor types:

- ATRT
- Brain stem glioma
- Choroid plexus tumors, choroid plexus carcinoma, choroid plexus papilloma
- Craniopharyngioma
- Cysts
- Desmoplastic infantile astrocytoma
- Ependymoma
- Germ cell tumors
- Medulloblastoma
- Neurofibromatosis
- Oligodendrogloma
- Optic glioma
- PNET

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 oncologists; pediatric radiation oncologists; 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 will 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 biopsy following surgery will help to classify and grade the tumor.

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

- Chemotherapy
- Conventional radiation therapy
- 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 the newest therapies while they are being developed. The ABTA can assist parents who want to better understand the risks and benefits of clinical trials, patient rights and protections in a trial and more. Contact the ABTA for more information on clinical trials, as well as other pediatric brain tumor resources.

The ABTA also offers a service that links brain tumor patients with appropriate clinical trials based on the patient's tumor type and treatment history. TrialConnect®, is a free and confidential service. For more information, call 877-769-4833 or go to [www.abtrialconnect.org](http://www.abtrialconnect.org).

### 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 effects later on in life from the treatments they have received. Access to detailed medical information will help your child 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 a wide range of support services 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

You don't 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. Children and teens have misconceptions that must be addressed, including:

- 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 “Coping” on pg 63.

### 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 requires brothers and sisters to be left in the care of other family or friends. Everyone is worried.

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 help you communicate effectively with family members, too.

#### 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. They want to visit the hospital, send cards, organize a bake sale. If that is the case, try to find ways for them to help. It will give them a concrete way to process their own emotions and

to demonstrate some small measure of control over what is happening in their lives.

### IMPACT ON RELATIONSHIPS

A crisis can bring out the best in relationships, but 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 and the like. 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” keeping all informed and updated.

#### ABTA CareLine

ABTA's Careline can be a family's single best resource. Health care professionals work with patients, families and caregivers daily to address a wide variety of needs. They are caring, compassionate and trained professionals who listen with their heart and respond with wisdom and resources accumulated over many years. They can help you connect with a wide range of support services available to you through the Internet and through community-based or institution-based resources. To connect with the ABTA, please call the CareLine at 800-886-ABTA (2282) or send us an email at [abtacares@abta.org](mailto:abtacares@abta.org).

### **ABTA Connections Online Support Community**

The American Brain Tumor Association

Connections online support community connects patients, families, friends and caregivers for support and inspiration. Unlike social media outlets such as Facebook or Twitter, 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 traversing a similar brain tumor journey.

Learn more at [www.abta.inspire.com](http://www.abta.inspire.com).

### **LATE EFFECTS OF TREATMENT**

Treatment decisions can have unintended consequences. These complications, known as “late effects” of treatment 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 them. Surgery, radiation therapy and chemotherapy can all contribute to late effect complications.

Late effects vary considerably based upon multiple factors including a person’s age, tumor type and location, treatment type and duration. Some effects may be apparent almost immediately, but some may not emerge until years after treatment has stopped. 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

- 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 son or daughter’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 your child’s specific 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.

For more information on near- and long-term effects of pediatric brain tumors and/or adolescent and young adult resources visit [www.abta.org](http://www.abta.org).

## 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 normalcy.

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 may have lots of 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, contact their school regarding your child’s diagnosis and treatment. Keep teachers updated. 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 and/or behavioral changes.

If your child has physical or learning disabilities following a brain tumor diagnosis and/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

There is ongoing research which shows that children treated for brain tumors may experience neuropsychological effects following treatment. Neuropsychological testing is done to help define the impact and identify learning disabilities. 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. Turn to the ABTA for help and additional resources.

For more information on pediatric brain tumors, please visit [www.abta.org](http://www.abta.org), call our CareLine at 800-886-ABTA (2282) or send us an email at [abtacares@abta.org](mailto:abtacares@abta.org).

## Chapter 9: Coping

Whether you are an individual with a brain tumor, a caregiver, family member or friend, you may still be trying to make sense out of the words “brain tumor.” You may be experiencing feelings of fear, uncertainty and 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 or become numb and hide or deny their feelings. Some people may refuse to discuss or even acknowledge their diagnosis. The 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. For patients, the thought “Why me?” is common. Life may feel very unfair. Some people develop a sense of resignation about living with the physical or emotional changes that may come with a brain tumor diagnosis. Others may feel resentment because they or a loved one has this disease and other people do not. For some people, this resentment may turn into depression.

A sense of acceptance will eventually set in as you realize that the brain tumor is a reality. Once 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.

It may sound strange, but some feel that their personal lives change for the better after a major diagnosis such as a brain tumor. It does make people re-evaluate 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 better and the next day feel uneasy again. Not everyone shows their emotions, nor does everyone experience the same

feelings. The important thing to remember is that we all experience a wide array of emotions and it all depends on how we acknowledge and cope with them.

### A Special Note on Anxiety and Depression

While being treated for a brain tumor, it is not uncommon for patients to experience anxiety or depression. These two common side effects should also be monitored and treated by your health care team as any other physical condition.

#### Anxiety

It is normal for people to experience anxiety when going through stressful times. Many people feel “anxious” while waiting for test results or when 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. It is important 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/she may suggest medication or an appointment with a psychiatrist, psychologist or social worker.

#### Depression

While depressed feelings can be normal, some people may become very depressed and need help in dealing with these feelings. Some of the symptoms of major depression are: persistent depression or no feelings whatsoever; irritability; loss of enjoyment and pleasure in people or activities that are normally enjoyable; difficulty sleeping – such as trouble falling asleep, waking too early, being unable to fall asleep again or sleeping too much; loss of appetite; or wanting to give up or to inflict self-harm. When these feelings persist for more than two weeks or when they are severe, it is important to bring the symptoms to the attention of a doctor. The 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 first be diagnosed.

### INCLUDING FAMILY AND FRIENDS

Life is about to change. For many, it helps to share your situation with family and friends who can offer support and help you work through your options. But 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 having a care conference or meeting with your doctor, health care 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. A family that understands your diagnosis and available treatment options has the opportunity to be supportive and helpful.

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, a meal on the day of your doctor visit, transportation 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, reach for that list. Or create an online calendar with tasks that need to be done.

Although many people will be supportive, there will be friends who simply find it difficult to deal with or even acknowledge your illness. Not knowing what to say, some people may avoid contact or conversation. Also, be prepared for well-meaning friends and neighbors who insist upon telling you stories about “miraculous” cures. Don’t 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 with whom you are comfortable and upon whom 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 seem bewildered as they take on responsibilities previously handled by the other person.

Every marriage survives by a set of “unwritten” rules that partners live by in their everyday lives. During the years of marriage, spouses take on certain roles, such as caretaker or provider within the family. They work out the labor issues for the household and establish guidelines for their intimate relationship and personal friendships. The diagnosis of a brain tumor can change those rules. Neither spouse may know exactly how to react to each other, especially if caretaker and provider roles have changed. Some couples find they start feeling uncomfortable with each other, not as close and sense they are losing the intimacy of their relationship. You may begin arguing or disagreeing with each other. At this moment, there is a temptation 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 major medical diagnosis is disruptive to a person's personal and family life, it can equally be an opportunity for renewal, hope and a new relationship. Spouses should talk about the changes in their 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, social worker or psychologist. A professional may be able to help with the adjustments and changes associated with the medical diagnosis and its effect on personal relationships.

Think about the ways you've handled previous crises in your life. Are you comfortable sharing your concerns? If so, to 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 now, too.

As you move further into the brain tumor experience, you'll begin to find resources you didn't 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 healthful meals, even when you're not very hungry. Moderate exercise (if approved by your doctor) can help fight fatigue, depression and improve your sleep. If you wish, allow yourself to feel unhappy – but realize there is help available to those whose sadness is consuming or excessive.

### TALKING WITH CHILDREN

If you are a parent with young children and you have a brain tumor, try to anticipate your children's 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 aren't easily answered; reply honestly and simply. Answer what is asked of you 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 – and don't worry about being an artist. Use their questions as a guide to the information you convey.

There are many books available that can help parents explain their illness to children. Read these books with your children; offer them the opportunity to ask questions and to 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..." or, "The doctor wants to do some tests to find out why you are 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 operate and take it out. The operation will help get rid of the headaches.”
- “A brain tumor is a collection of abnormal ‘cells’ in the brain which 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.

It also is important for parents to 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 easy to predict, are consistent and make them feel safe. Therefore, it is important that parents agree not only on what to tell their children, but 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’ve heard from friends or personal experience with grandparents or neighbors. Give older children the opportunity to ask questions or share their concerns. “Is there anything you’d like to ask?” can be difficult to say, but can open the door to clarifying misinformation as well as providing reassurance.

Some children, especially pre-teens and teenagers, may begin to act out. They may get in trouble, begin to have problems in school or begin to do things they normally would not do. This is usually a sign that the child is having trouble coming to grips with what is happening within the family. It may be time for more direct action. 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, it is time to 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 of love. 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 won’t be the same without their parent, the child will know that mom or dad cared enough to ensure that someone would be there.

The ABTA can help with sample conversations; support groups for parents, children and siblings; and other information. Contact the ABTA CareLine at 800-886-ABTA (2282) or [abtacares@abta.org](mailto:abtacares@abta.org).

## REDUCING STRESS

For most people, fears of the unknown and an uncertain 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, permit 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 her/his 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 crisis. Close friends, religious leaders or your health care 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. Many community programs are available – learn what they are 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'll need information on all your options.
- Ask your doctors this question: “What questions should I be asking that I don't know I should be asking?”
- Don't 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'll save your energy.
- Take advantage of help offered to you. Family, friends and neighbors can make life easier, if you let them. And don't worry about repaying the favors.

- Allow yourself to cry. It's 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's 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 don't 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 there is often a need to connect with others in similar situations. There are many ways to reach for support but not all types of support are appropriate for everyone. We offer some information and resources to help patients and families decide which support resources might be best for them.

### 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 one another 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 may provide a comfortable, safe forum 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 are strictly supportive, while others are strictly educational; many groups are both supportive and educational.

Groups also are either "open" or "closed" to new members. Most groups are "open" and ongoing, meaning that members may join or leave the group as their needs change. There can be much stability

in these groups. However, as members come and go, the personality of the group may continually change. In contrast, time-limited “closed” groups meet for a fixed period of time – for example, six to eight weeks – with the same individuals. Once the group starts, it is closed to new members until the end of the specified time period.

All groups, whether open or closed, ongoing or time-limited, 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.

The major advantage of a face-to-face support group is the warmth and closeness that often develops when people, who are typically living in the same community or area, interact with and support each other on a very personal level. 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 doesn’t 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 the support group closest to you, contact the ABTA CareLine at 800-886-ABTA (2282) or send an email to [abtacares@abta.org](mailto:abtacares@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 important 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 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. And 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’s best to only share information that you are completely comfortable sharing with strangers.

**ABTA Connections Online Support Community**

The American Brain Tumor Association Connections online support community connects patients, families, friends and caregivers for support and inspiration. Unlike social media outlets such as Facebook or Twitter, 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 traversing 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.org/inspire.com](http://www.abta.org/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/or family counseling when more formal assistance is needed. For help in locating a professional counselor or therapist, please contact the ABTA CareLine at 800-886-ABTA (2282) or send an e-mail to [abtacares@abta.org](mailto:abtacares@abta.org).

**Neuropsychologist**

A neuropsychologist is a mental health professional with expertise in assessing and treating problems of psychological functions and behavior, as they relate 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. A neurologist or neurosurgeon can recommend a board certified neuropsychologist or you can contact the ABTA for more information at 800-886-ABTA (2282) or send an email to [abtacares@abta.org](mailto:abtacares@abta.org).

**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. It is a time when it can be difficult to do nothing after having done so much.

Your task now is becoming 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 health care team. Learn about healthful 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, even if it's somewhere in the past. Learn where it belongs and what feels comfortable for your family. Your priorities will also change with time. That's 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 website, [www.abta.org](http://www.abta.org), offers extensive brain tumor information, treatment and research updates, lists of family and professional brain tumor meetings and conferences, and information about upcoming ABTA events.

## Chapter 10: For the Caregiver

When someone we love is ill, we want to do everything we can to help them. Being a caregiver to a family member or close friend can be one of the most rewarding experiences a person can have. It also can 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. That can mean 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

The words *brain tumor* can be overwhelming. It's common for families to make a visit to the doctor, hear terms and phrases they've never heard before and then be asked to make a decision. Back at home, there may be a flood of doubts as to what was heard and whether it was understood. If you have questions about the information your family was given, call the doctor. If you did not understand something that was told to you or you have additional questions you forgot to ask, call the doctor. The doctor believes you understand everything that was said unless you speak up. And, having answers to your questions can be a great stress reducer.

## ASSESS WHAT YOU NEED

Have a clear sense of what it is that would be most helpful to you as a 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 the grocery shopping or someone to come in 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

Don't do everything yourself if there are 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 will be other tasks which also need attention. Look beyond the 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 and, thus, 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, baby-sit 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 also can suggest they help in some less tangible 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 also is available in many communities.

## UTILIZE ONLINE SUPPORT GROUPS AND WEBSITES

There are a variety of Internet-based resources, including online support groups, listservs, chat rooms, message boards and other Internet communities of interest, through which caregivers can share information and support 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 by the Internet. Most online forums require that you subscribe or register prior to sending messages. After subscribing as directed, you'll receive a confirmation as well as a welcome message. Each forum has its own etiquette rules and procedures. Save those guidelines; they will also tell you how to unsubscribe should you so choose. The group may also provide you with directions for obtaining your email in digest form, a simple way of gathering all the participant messages into one email or one folder.

#### **ABTA Connections Online Support Community**

The American Brain Tumor Association Connections online support community connects patients, families, friends and caregivers for support and inspiration. Learn more at [www.abta.inspire.com](http://www.abta.inspire.com).

#### **BE KIND TO YOURSELF**

When 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?" When we apply this test, I think most of us have no problem letting our hypothetical friend "off the hook." 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**

It is not uncommon for caregivers to neglect their own health while trying to provide the best care possible for the person they are caring for. And yet neglecting your health can leave you vulnerable to exhaustion and disease. To prevent illness, 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 also can be very helpful. A relative, a friend or a member of your clergy all can be important parts of your social network. Sometimes it helps to let that person know that you don't 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 not to take their advice. And make sure you feel better after talking with the person you select. Support that doesn't feel like support usually isn't.

Support groups with other caregivers can be another valuable resource. Many of these groups welcome family members and caregivers. We can provide you with a list of support groups in your state. Call the ABTA CareLine at 800-886-ABTA (2282) or email [abtacares@abta.org](mailto:abtacares@abta.org).

If you find that friends and family and other non-professional support resources just aren't enough or you continue to feel consistently overwhelmed, you may wish to consult with a mental health professional. Your doctor can refer you to one. Professional mental health associations also can help you locate someone in your area.

#### **MAKE TIME FOR YOURSELF**

Try to take a break from caregiving for at least a few hours or longer each week, if you can. It can

be difficult to leave your loved one, especially if they are very ill, but it may be the most important thing you do for that person and for yourself. Taking this time is not being selfish. Rather, it 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 find relaxing. Meditation, guided imagery and exercise or movement classes may 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 responsibility and worry.

### 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 “out there winging it.” Don’t 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 an immaculate house, may not be important now. And some things that weren’t 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’ve 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’s amazing how much better you feel being able to cross it off your mental “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 don’t have to go through this journey alone. The American Brain Tumor Association is here to help.

Visit us at [www.abta.org](http://www.abta.org) to find additional brochures, read about research and treatment updates, connect with a support community, join a local event and more.

We can help you better understand brain tumors, treatment options, and support resources. Our team of health care professionals are available via email at [abtacares@abta.org](mailto:abtacares@abta.org) or via our toll-free CareLine at 800-886-ABTA (2282).

## Chapter 11: Brain Tumor Facts and Statistics

The facts and statistics here include brain and central nervous system tumors (including spinal cord, pituitary and pineal gland tumors). We continually update these statistics, as they become available, on our website, [www.abta.org](http://www.abta.org). This material was last updated in 2014. We thank the Central Brain Tumor Registry of the United States (CBTRUS) for their assistance with that update.

These numbers address incidence, trends and patterns in the United States only. For more information, please visit CBTRUS at [www.cbtrus.org](http://www.cbtrus.org).

## 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 (males and females) under age 20 (leukemia is the first)
- Second leading cause of cancer-related deaths in males ages 20–39 (leukemia is the first)
- Fifth leading cause of cancer-related deaths in females ages 20–39

### Incidence Statistics

An estimated 68,470 new cases of primary brain tumors are expected to be diagnosed in 2015 and includes both malignant (23,180) and non-malignant (45,300) brain tumors. These estimates are based on an application of age-sex-race-specific incidence rates from the 2014 CBTRUS Statistical Report using 2007-2011 SEER and NPCR data to project 2015 US population estimates for the respective age-sex-race groups (estimation methodology can be found at [www.idph.state.il.us/cancer/statistics.htm#PR](http://www.idph.state.il.us/cancer/statistics.htm#PR)).

In 2015, approximately 4,620 children between the ages of 0-19 will be diagnosed with primary brain tumors. In 2015, an estimated 3,420 new cases of childhood primary brain and central nervous system tumors are expected to be diagnosed in children ages 0 – 14.

### Prevalence Statistics

It is estimated that during the year 2010 more than 688,096 people in the United States were living with the diagnosis of a primary brain or central nervous system tumor. Specifically,

more than 138,054 persons were living with a malignant tumor and more than 550,042 persons were living with a non-malignant tumor.

## TUMOR-SPECIFIC STATISTICS

- *Meningiomas represent 36% 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 28% of all brain tumors and 80% of all malignant tumors.*
- *Glioblastomas represent 17% of all primary brain tumors and 54% of all gliomas.*
- *Astrocytomas and glioblastomas combined represent 76% of all gliomas*
- *Nerve sheath tumors (such as acoustic neuromas) represent about 8% of all primary brain tumors.*
- *Pituitary tumors represent 13% of all primary brain tumors.*
- *Lymphomas represent 2% of all primary brain tumors.*
- *Oligodendrogiomas represent 2% of all primary brain tumors.*
- *Medulloblastomas/embryonal/primitive tumors represent 3% of all primary brain tumors.*
- *The majority of primary tumors (36%) are located within the meninges, followed by those located within 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.*

*The cancers that most commonly metastasize to the brain are lung and breast.*

## 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).

### **Brain Stem**

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

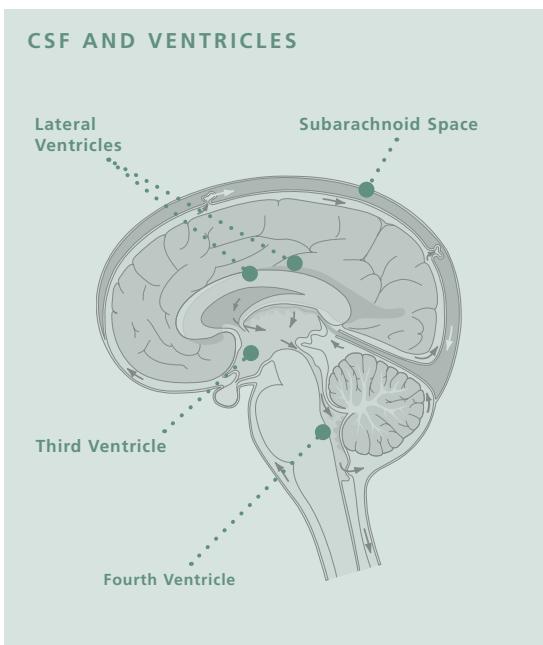
### **Cerebrospinal Fluid (CSF)**

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 largest area of the brain is the cerebrum, which consists of the right and left cerebral hemispheres. In general, 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.

#### **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**

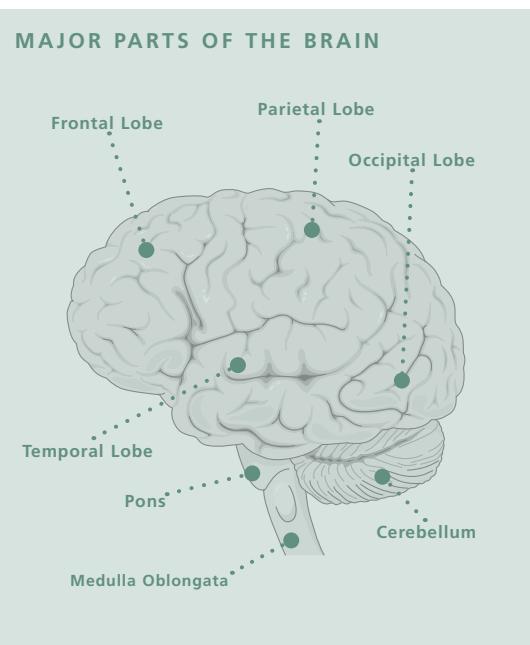
The corpus callosum is made of nerve fibers, deep in the brain, that connect the two halves of the cerebral hemispheres.

#### **Cranial Nerves**

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

#### **Glia**

Glia, the supportive tissue of the brain, is made up of glial cells. The most common glial cells are astrocytes and oligodendrocytes. Ependymal cells are another form of glia.



Glia cells are the origin of the largest percentage of brain tumors, i.e., astrocytomas (including glioblastoma), oligodendrogiomas 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**

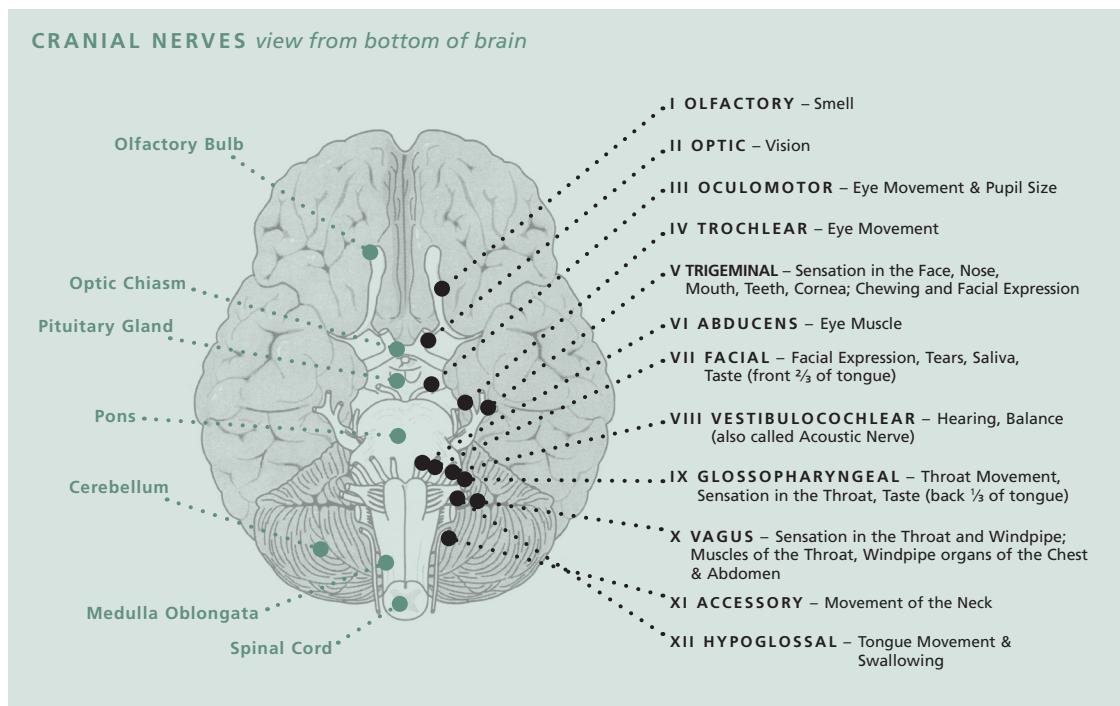
The hypothalamus regulates sleep cycles, body temperature and other metabolic processes. It acts like an endocrine gland by producing hormones and sending messages that control the hormonal secretions of the pituitary gland.

#### **Medulla Oblongata**

The medulla oblongata, a part of the brain stem, 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

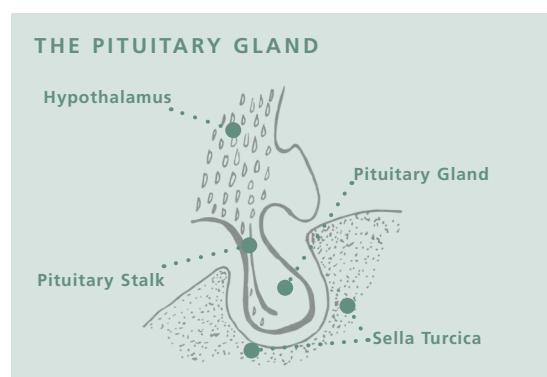
pituitary produces several hormones including prolactin, corticotropin and growth hormone.

#### Pons

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

#### Posterior Fossa (Infratentorium)

The tentorium separates the posterior fossa from the cerebral hemispheres. The area below the tentorium is called the infratentorium or the posterior fossa. This area within the skull contains the cerebellum and the brain stem. The area above the tentorium is called the supratentorium.



### Reticular Formation

The reticular formation is the central core of the brain stem. It connects with all parts of the brain and brain stem.

### 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) of the brain 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.

### Supratentorium

The supratentorium is the area above the tentorium containing the cerebral hemispheres.

### Tentorium

The tentorium is a flap of meninges separating the cerebral hemispheres from the structures in the posterior fossa.

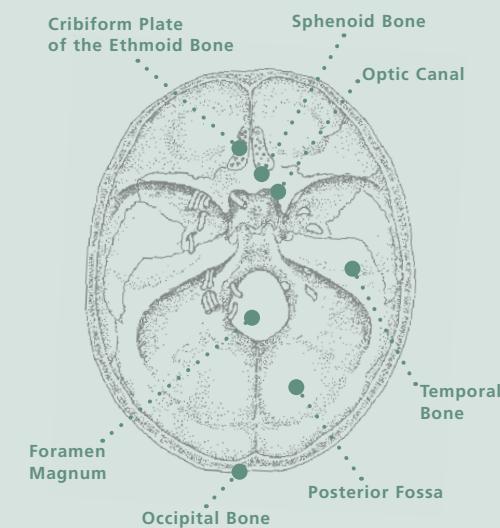
### Thalamus

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

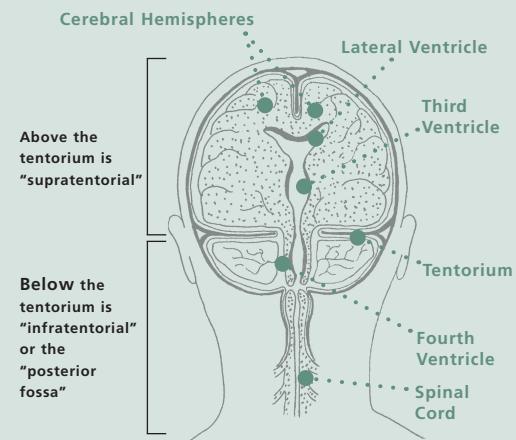
### Ventricles

These are connected cavities (the lateral, third and fourth ventricles) 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.

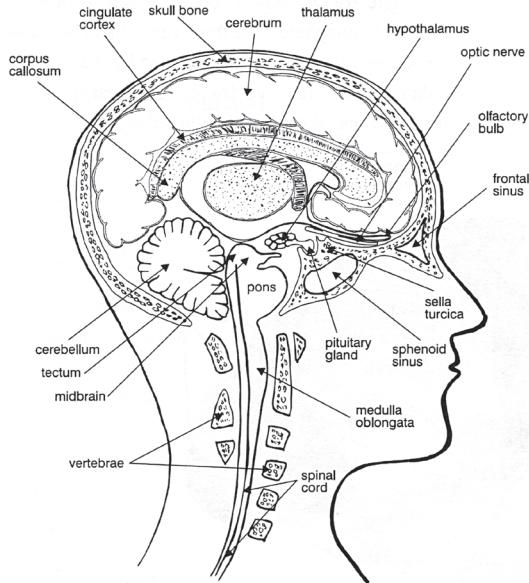
### SKULL BASE



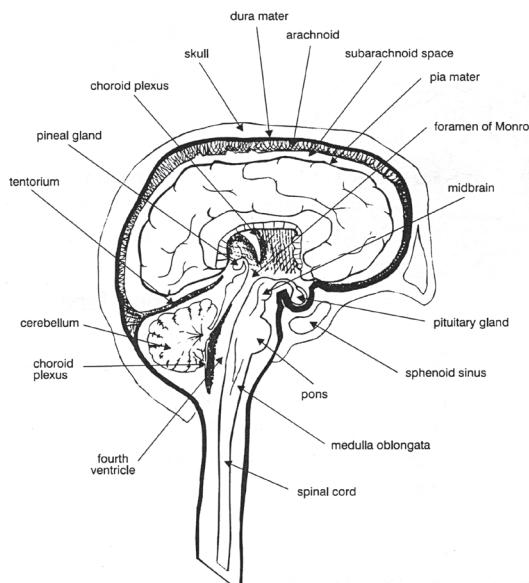
### THE TENTORIUM



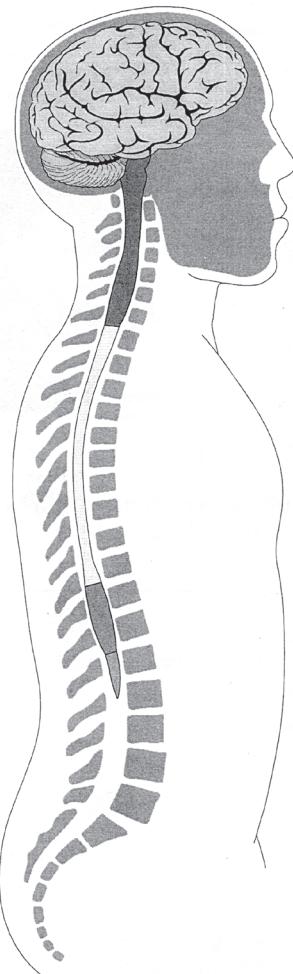
There are two lateral ventricles, one in each cerebral hemisphere. The third ventricle is beneath the corpus callosum and surrounded by the thalamus. The fourth ventricle is an expansion of the central canal of the medulla oblongata.



CROSS SECTION OF THE BRAIN

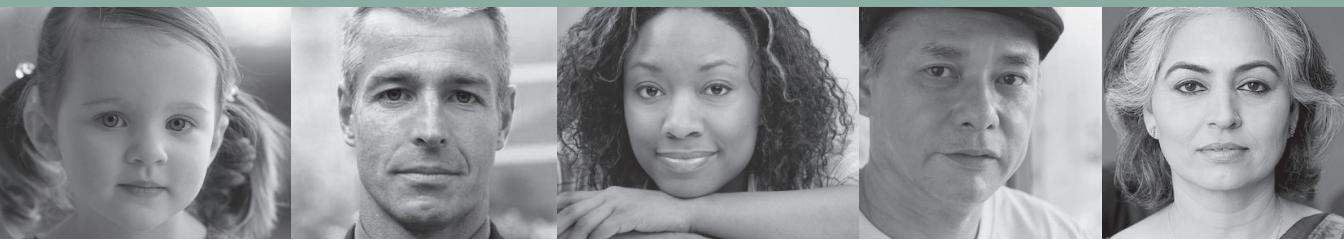


SIDE VIEW OF THE BRAIN



CENTRAL NERVOUS SYSTEM

# Providing and Pursing Answers<sup>TM</sup>



The American Brain Tumor Association (ABTA) is a trusted resource for brain tumor patients, caregivers and health care professionals around the country. From a wide range of publications to webinars and educational programs, our goal is to ensure that you have the tools and information you need. Visit our web site at [www.abta.org](http://www.abta.org) to see a full scope of all that we provide. Our complete line of patient education publications can be downloaded from our web site, and hard copies can be requested by calling our CareLine at 800-886-ABTA (2282).

## AMERICAN BRAIN TUMOR ASSOCIATION PUBLICATIONS AND SERVICES

### CARE & SUPPORT

CareLine: 800-886-ABTA (2282)

Email: [abtacares@abta.org](mailto:abtacares@abta.org)

### PUBLICATIONS

*About Brain Tumors: A Primer for Patients and Caregivers*

*Brain Tumor Dictionary*

#### Tumor Types:

Ependymoma

Glioblastoma and Malignant Astrocytoma

Medulloblastoma

Meningioma

Metastatic Brain Tumors

Oligodendrogioma and Oligoastrocytoma

Pituitary Tumors

#### Treatments:

Chemotherapy

Clinical Trials

Conventional Radiation Therapy

Proton Therapy

Stereotactic Radiosurgery\*

Steroids

Surgery

All publications are available for download in Spanish.  
(exception is marked \*)

### CLINICAL TRIALS

TrialConnect®: [www.abtatrialconnect.org](http://www.abtatrialconnect.org) or  
877-769-4833

*More brain tumor resources and information  
are available at [www.abta.org](http://www.abta.org).*



American  
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## NOTES/QUESTIONS

## AMERICAN BRAIN TUMOR ASSOCIATION

To find out how you can get more involved locally, contact [volunteer@abta.org](mailto:volunteer@abta.org) or call 800-886-1281

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**For more information contact:**

CareLine: 800-886-ABTA (2282)

Email: [info@abta.org](mailto:info@abta.org)

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

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