Brain Tumor Primer

A COMPREHENSIVE INTRODUCTION TO BRAIN TUMORS, 9TH EDITION

American Brain Tumor Association
A Word About ABTA

Founded in 1973, the American Brain Tumor Association was the first national, non-profit organization dedicated solely to brain tumors. Today, ABTA is a leader in funding brain tumor research, and providing patient-family resources and support.

We gratefully acknowledge the following for their assistance in the preparation of this edition: Terri S. Armstrong, PhD, ANP-BC, Associate Professor Department of Integrative Nursing Care, University of Texas School of Nursing, Houston, Texas; Jill Barnholtz-Sloan, PhD, Case Comprehensive Cancer Center, Case Western Reserve University School of Medicine, Cleveland, Ohio; Steven S. Brem, MD, Chair, Department of Neuro-Oncology, H. Lee Moffitt Cancer Center & Research Institute, Tampa, Florida; Susan Chang, MD, Director, Division of Neuro-Oncology, University of California San Francisco, California; Mark R. Gilbert, MD, Professor, Department of Neuro-Oncology, Division of Cancer Medicine, The University of Texas M. D. Anderson Cancer Center, Houston, Texas; Bridget McCarthy, PhD, Central Brain Tumor Registry of the United States, Hinsdale, Illinois; Sridhar Nimmagadda, PhD, Assistant Professor of Radiology, Medicine and Oncology, Johns Hopkins University, Baltimore, Maryland; Mady Stovall, RN, MSN, NP, Neuro-Oncology Nurse Practitioner, Kaiser Permanente, Redwood City, California; Michael Taylor, MD, PhD; Assistant Professor, Departments of Surgery and Laboratory Medicine and Pathobiology, University of Toronto, Hospital for Sick Children, Toronto, Ontario, Canada; Michael A. Vogelbaum MD, PhD, Associate Director, Brain Tumor and Neuro-Oncology Center, Director, Center for Translational Therapeutics, Cleveland Clinic, Cleveland, Ohio; Vicky Holetz Whittemore, PhD, Vice President & Chief Scientific Officer, Tuberous Sclerosis Alliance, Silver Spring, Maryland. We also thank Gail Segal for her tireless contributions to the early versions of this publication.

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ISBN 0-0944093-88-4
Dedication

This Primer is dedicated to the memory of Jerome Braverman and Florence Braverman.

Jerome served as president of the Association from 1979 to 1981 and his efforts made the first edition of this Primer possible. Active in a variety of civic and charitable causes, Jerry often went out of his way to help those in need. His broad shoulders, patient ear, and soothing voice are missed.

Florence devoted herself to responding personally with notes of thanks to the thousands of people who supported the Association’s work with their contributions and good wishes. A caring and generous person, the loss of Florence’s kindness and understanding leaves a void that cannot be filled.

This printing was made possible by a generous donation from Genentech, the Byrne Foundation, and the Butler Family Foundation.
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.

The American Brain Tumor Association wrote this book to help you, your family and your friends learn more about brain tumors. We hope this knowledge will offer a degree of comfort, and help you feel more in control of your life during this difficult time.

Also know that the American Brain Tumor Association is here to help you throughout this journey. Our team of compassionate, licensed Patient Services professionals can provide additional information about tumors, treatment, and support resources. We also urge you to visit our Web site at www.abta.org, and consider joining our online community, Connections (https://connections.abta.org) where you’ll find others just like you who are interested in sharing their experiences, information and support.

Please call us at 800-886-2282, or e-mail us at info@abta.org.
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The material contained in this book is provided for informational purposes only and should not be considered as medical advice. Inclusion does not imply the American Brain Tumor Association’s endorsement or recommendation. Always check with your doctor about your specific medical problem or condition. Information herein is subject to change.
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Publications & Services … Inside Back Cover

Please note that the majority of these chapters print in two columns. If you do not see two columns when reading the information please contact the American Brain Tumor Association at 800-886-2282. We can provide an alternative format for you. This publication is also available on our website at www.abta.org.
Living creatures are made up of cells. 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 and understanding, 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 brain and spine will help you understand the symptoms of brain tumors, how they are diagnosed, and how they are treated.
**Terminology**

Detailed, enlarged diagrams of the brain can be found on pages 7 – 8.

**BASAL GANGLIA**
The basal ganglia are masses of nerve cells deep within the cerebral hemispheres.

**BRAIN STEM**
The brain stem is the bottom-most portion of the brain, connecting the cerebrum with the spinal cord. The midbrain, pons, medulla oblongata and reticular formation are all part of the brain stem.

**CEREBELLOPONTINE ANGLE**
The angle between the pons and the cerebellum.

**CEREBELLM**
The cerebellum is the second largest area of the brain. It consists of two hemispheres, or halves, as well as a middle portion. The cerebellum is connected to the brain stem.

**CEREBROSPINAL FLUID (CSF)**
CSF is the clear, watery fluid made in the ventricles that bathes and cushions the brain and spinal cord. It circulates through the ventricles and the subarachnoid space.

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, which is nerve cells. Nerve cells control brain activity.

The inner portion of the cerebrum is mostly white matter with nerve fibers called axons. These are insulated by a fatty substance known as myelin. White matter carries information between nerve cells by conducting electrical impulses.

**CSF AND VENTRICLES**

**CEREBRUM/CEREBRAL HEMISPHERES**
The largest area of the brain is the cerebrum which consists of the right and left 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.

**CHOROID PLEXUS**
The choroid plexus produces spinal fluid that flows through the ventricles and meninges 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**
There are 12 pairs of cranial nerves. Their functions are described in the illustration on the following page.

**GLIAL TISSUE (NEUROGLIA)**
Glia is the supportive tissue of the brain. The cells which make up this tissue are called glial cells. The most common glial cells are astrocytes and oligodendrocytes. Ependymal cells are another form of glia.
Glial cells are the origin of the largest percentage of brain tumors, i.e., astrocytomas (including glioblastoma), oligodendrogliomas, and ependymomas. Astrocytes are involved with the blood brain barrier and brain metabolism. Oligodendrocytes maintain the myelin covering of nerve cells. Myelin helps transmit information between nerve cells.

**HYPOTHALAMUS**
The hypothalamus makes up part of the wall of the third ventricle and is the base of the optic chiasm.

**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. A tumor called meningioma arises from the meninges.

**MIDBRAIN**
The midbrain is the short portion of the brain stem between the pons and the cerebral hemispheres. The top of the midbrain is called the tectum (or tectal area). 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. It produces the hormone melatonin. This hormone is believed to control the biological rhythm of the body.

**PITUITARY GLAND**
The pituitary gland is attached to, and receives messages from, the hypothalamus. The pituitary gland is composed of two lobes — the anterior and the posterior. Several hormones are produced by the pituitary including prolactin, corticotropin, and growth hormone.
**PONS**
The pons, a part of the brain stem, contains the origins of the 5th, 6th, 7th, and 8th cranial nerves.

**POSTERIOR FOSSA (INFRAVENTORIUM)**
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. The sella turcica is a hollow in the skull bone that contains the pituitary gland.

**SKULL BASE**
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, i.e., nerve fibers. It begins in the medulla oblongata 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. Cerebrospinal fluid flows through the meninges.

**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.

**THE TENTORIUM**

![Diagram of the tentorium and related structures](image)

**THALAMUS**
The thalamus surrounds the third ventricle.

**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 subarachnoid space of the meninges.

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

- Thalamus
- Hypothalamus
- Optic nerve
- Olfactory bulb
- Frontal sinus
- Pituitary gland
- Sella turcica
- Sphenoid sinus
- Pons
- Medulla oblongata
- Cerebellum
- Corpus callosum
- Cingulate cortex
- Visual cortex
- Skull bone
- Cerebrum
- Vermis
- Tectum
- Midbrain
- Spinal cord
- Vertebrae
- Spinal cord
s I d e  v I e W  o f  t h e  B r a I n

f o u r t h  v e n t r I c l e

p I t u I ta r y  g l a n d

s p I n a l  c o r d

c h o r o I d  p l e x u s

c e r e B e l u m

t e n t o r I u m

p I n e a l  g l a n d

m e d u l l a  o B l o n g ata

p o n s

s p h e n o I d  s I n u s

m I d B r a I n

f o r a m e n  o f  m o n r o

c h o r o I d  p l e x u s

s k u l l

d u r a  m at e r

a r a c h n o I d

s u B a r a c h n o I d  s pa c e

p I a  m at e r

S I D E  V I E W  O F  T H E  B R A I N
The adult body normally forms new cells only when they are needed to replace old or damaged ones. Infants and children form 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. 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 the pathological features contribute to the outcome.
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 can be 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, the 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 to be life-threatening — although the tumor and its cells would not be classified as malignant.

Malignant Brain Tumors

A malignant brain tumor is usually rapidly 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.

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.”

Cancer is a disease defined by:

- unregulated growth of abnormal cells
- abnormal cells that grow into/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 or
- are located in a critical part of the brain or
- 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.
**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, to plan treatment, and to predict outcome. 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 of neurons—for example, gangliocytoma and ganglioglioma—are examples of 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.

Grade IV 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.

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. A benign growth might become malignant. In some tumors, a lower-grade tumor might recur as a higher-grade tumor. Your doctor can tell you if your tumor might have this potential.
**Change of Diagnosis**

Although it may initially seem alarming, your diagnosis and the name of your tumor might be changed. 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.

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**Tumor Staging**

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

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

Staging information often influences treatment recommendations and prognosis.

---

**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 type of therapy is also instrumental. Certain tumors, although malignant, can be cured by radiation therapy or chemotherapy. Others, by virtue of their location, may ultimately be lethal in spite of their “benign” appearance under the microscope.

Additional information about tumor prognosis is also available in our Focusing on Tumors series of publications.

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**About “Lesions”**

“Lesion” is a general term which refers to any change 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 s/he believes this to be. Treatment will be determined based on the type of lesion.
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 in children.

Brain tumors are:

- the second leading cause of cancer-related deaths in children under age 20 (leukemia is the first)
- the second leading cause of cancer-related deaths in males up to age 39
- the second leading cause of cancer-related deaths in females under age 20.
- the fifth leading cause of cancer-related deaths in females ages 20–39.6

The facts and statistics in this chapter include incidence, trends, and patterns in the United States only. We update these statistics at our web site—www.abta.org—as they become available.

We express our appreciation to the Central Brain Tumor Registry of the United States (CBTRUS) for their efforts in preparing many of these updates. For more information, please visit CBTRUS at www.cbtrus.org.
Incidence Statistics

An estimated 62,930 new cases of primary brain tumors are expected to be diagnosed in 2010 and includes both malignant (23,720) and non-malignant (39,210) brain tumors.

These estimates are based on an application of age-sex-race-specific incidence rates from the 2010 CBTRUS Statistical Report using 2004 – 2006 SEER and NPCR data to projected 2010 US population estimates for the respective age-sex-race groups (estimation methodology can be found at http://www.idph.state.il.us/cancer/statistics.htm#PR).

Incidence is the number of people newly diagnosed in one year. Rate is the measure of the amount of a disease in a specific population. It is calculated by counting the number of people with the disease and dividing by the total population at risk.

In 2010, approximately 4,030 children younger than age 20 will be diagnosed with primary brain tumors, of which 2,880 will be under age 15.

Brain tumors are the most common of the solid tumors in children, and the leading cause of death from solid tumors. Brain tumors are the second most frequent malignancy of childhood; leukemia is the most common.

Although statistics for brain metastases are not readily available, it is estimated that more than 150,000 cancer patients per year will have symptoms due to a metastatic brain tumor or a metastatic brain tumor in the spinal cord. Metastatic brain tumors begin as a cancer elsewhere in the body and spread, or metastasize, to the brain. Primary brain tumors are tumors that begin in the brain and tend to stay in the brain.

Regarding Incidence Trends

The incidence of all primary brain and central nervous system tumors appears to increase steadily with age. The lowest incidence rate is among children less than 20 years (4.7 per 100,000 person years). The rate increases steadily until age 75 – 84, when it peaks at 65.5 per 100,000 person years. After age 85, the incidence rate drops to 64.2.

Prevalence Statistics

It is estimated that during the year 2004 more than 612,000 people in the United States were living with the diagnosis of a primary brain or central nervous system tumor. Specifically, more than 124,000 persons were living with a malignant tumor and more than 488,000 persons were living with a non-malignant tumor.

For every 100,000 people in the United States, approximately 209 are living following the diagnosis of a brain tumor. This represents a prevalence rate of 209.0 per 100,000 person years.

Prevalence is the number of people living with a disease at a given point in time.

The prevalence rate for all pediatric (ages 0 – 19) primary brain and central nervous system tumors was estimated at 35.4 per 100,000 with more than 28,000 children estimated to be living with this diagnosis in the United States in 2004.
**Pediatric Statistics**

An estimated 4,030 children under age 20 are expected to be diagnosed with a primary benign or malignant brain tumor in 2010.1 Of these, 2,880 will be less than 15 years of age, and 1,150 between the ages of 15 and 19.

The pediatric incidence rate of 4.71 per 100,000 person years is slightly higher in boys (4.75 per 100,000) than girls (4.66 per 100,000).1 Brain tumors are the second most frequent malignancy of childhood6 and the most common of the solid tumors in children.2 Brain tumors are the second leading cause of cancer-related deaths in children under the age of 20.6 Leukemia remains the first.2, 6

The majority of childhood tumors (17.2%) are located within the frontal, temporal, parietal, and occipital lobes of the brain. Tumors located in the cerebrum, ventricle, brain stem and cerebellum account for 6%, 6%, 11%, and 16% of all childhood tumors, respectively. Tumors located in overlapping or ‘other’ brain locations account for 14% of all childhood tumors.1

Gliomas account for a significant percentage of childhood tumors:

- 55% of all tumors and 71% of malignant tumors in children age 0 – 14
- 40% of all tumors and 74% of malignant tumors in children age 15 – 19.1

Trends in incidence of primary malignant brain tumors for children in the United States using Surveillance, Epidemiology, and End Results (SEER) Program data and a sophisticated statistical technique were evaluated in 1998.7 SEER is a program of the National Cancer Institute. It collects and analyzes information on cancer incidence, mortality, and survival in the U.S. SEER data does not include benign brain tumors. The incidence of brain malignancies did not increase steadily from 1978 to 1994 as previously reported, but rather “jumped” to a steady, higher rate after 1984 – 85. The timing of the “jump” coincided with the wider availability of magnetic resonance imaging (MRI) in the United States.

This finding, combined with the absence of any “jump” in corresponding mortality for the same period, appears due to improved diagnosis and reporting during the 1980s.

**Age-, Gender-, and Race-Specific Statistics**

The incidence rate of primary non-malignant and malignant brain and central nervous system tumors is 18.71 cases per 100,000 person-years. For all primary brain and other nervous system tumors, the incidence rate is 17.44 per 100,000 for males and 19.88 per 100,000 for females.1 Rates are age-adjusted to the year 2000 U.S. standard population.

Brain tumors are the:

- the second leading cause of cancer-related deaths in children under age 20
- the second leading cause of cancer-related deaths in males up to age 39
- the second leading cause of cancer-related deaths in females under age 20.
- the fifth leading cause of cancer-related deaths in females ages 20–39.6

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![% INCIDENCE OF GLIOMA BY AGE IN RELATION TO ALL OTHER BRAIN TUMORS](image-url)
Rates for all primary brain tumors combined are higher among Whites (18.89 per 100,000 persons) than African-Americans (17.14 per 100,000). The difference between these rates is statistically significant.

The overall incidence rate for primary brain and central nervous system tumors among Hispanics is 17.73 per 100,000, compared to 17.36 per 100,000 for non-Hispanic African-Americans and 19.15 per 100,000 for White non-Hispanics.

**Tumor-Specific Statistics**

Meningiomas represent 34% 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 32% of all brain tumors and 80% of all malignant tumors.

Glioblastomas represent 17% of all primary brain tumors, and 54% of all gliomas.

Astrocytomas represent 7% of all primary brain tumors.

Astrocytomas and glioblastomas combined represent 76% of all gliomas.

Nerve sheath tumors (such as acoustic neuromas) represent about 9% of all primary brain tumors.

Pituitary tumors represent 13% of all primary brain tumors.

Lymphomas represent 2% of all primary brain tumors.

Oligodendrogliomas represent 2% of all primary brain tumors.

Medulloblastomas/embryonal/primitive tumors represent 1% of all primary brain tumors.

The majority of primary tumors (33%) are located within the meninges, followed by those located within the frontal, temporal, parietal and occipital lobes of the brain (23%).

Metastatic brain tumors are the most common brain tumor, with an annual incidence more than four times greater than that of primary brain tumors.
The cancers that most commonly metastasize to the brain are lung and breast.

**Survival Trends**

In 2008, the American Cancer Society reported a significant decrease in the number of brain and central nervous system cancer deaths over the past 13 years. Deaths due to malignant brain tumors decreased 14.36% between 1991 and 2004.1

In an analysis of SEER data from 1973 – 2001, five year survival rates for those with malignant brain tumors showed improvement over a three decade period: 21% in the 1970’s, 27% in the 1980’s, and 31% in the 1990’s.1

SEER data from 1995-2006 shows a 34% survival rate for males and 37% rate for females.1

Children, age 0 to 19, had the highest five-year survival rate at 72% between 1995 and 2006. That survival rate diminishes as age increases, down to 5% for persons age 75 and older.1

For Whites, the five-year survival rate jumped from 22% between 1974 and 1976, to 34% between 1996 and 2003.1 For African Americans, the five-year survival rates for the same time periods increased from 27 to 37%.1

NOTE – The term “five year survival” does not mean that group of people lived only five years after the start of the study. It means the study followed them for only five years. Five years is a standard “goal” in measuring survival for most diseases.

Five year, or even ten year, survival statistics do not tell us how many people lived longer than the five or ten years of the study. Those statistics require longer-term follow-up of people diagnosed with the given disease, which can be challenging to do in our mobile society. It can be very difficult for researchers to stay in contact with patients for more than five or ten years given the frequency of American family moves.
Sources


For Additional Information

In 1990, the American Brain Tumor Association conducted a feasibility study to evaluate the status of brain tumor data collection, and to determine the practicality of starting a registry whose purpose would be the collection of statistics for both benign and malignant brain tumors. The results of that study highlighted both the need and feasibility of such a registry. The American Brain Tumor Association then incorporated the Central Brain Tumor Registry of the United States (CBTRUS), and provided organization and financial support to the new entity.

CBTRUS was incorporated as a not-for-profit organization in 1992 to provide a resource for the gathering and circulating of current information on all primary brain tumors, benign and malignant, for the purposes of:

- describing incidence and survival patterns
- evaluating diagnosis and treatment
- facilitating etiologic (causation) studies
- establishing awareness of the disease
- and, ultimately, for the prevention of all brain tumors.

State or regional tumor registries obtain information about brain tumor patients from hospitals in their area. CBTRUS began by collection information from four registries that were already collecting data on benign and malignant brain tumors. Using their preliminary data, CBTRUS conducted studies to determine diagnostic accuracy and data completeness. With the passage of the Benign Brain Tumor Cancer Registries Amendment Act (Public Law 107-260), government funded surveillance organizations in the US are required to collect data on all primary non-malignant, as well as malignant, brain and CNS tumors beginning in 2004. The data collected is used to define incidence roles of all primary brain tumors, and can be used by researchers to identify geographic clusters of patients.

Please visit the Web site of the Central Brain Tumor Registry at www.cbtrus.org. For more information or additional statistical data on primary brain tumors, contact CBTRUS at 244 East Ogden Ave, Suite 116, Hinsdale, IL 60521. Phone 630-655-4786, Email: cbtrus@aol.com.
Causes & Risk Factors

When someone learns 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 to my family,” and “how close are we to preventing this” are all normal questions.

Scientists trained in studying groups of people with the same disease are called “epidemiologists.” 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. Or, these genetic mutations/susceptibilities may 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. However, there are many scientific groups throughout the world focused on discovering the causes and/or risk factors for brain tumors.

**Environmental Factors**

Many studies have looked at a wide spectrum of environmental factors as possible causes for brain tumors. Of the long list of factors studied, only exposure to ionizing radiation has been consistently associated with 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 that used in medicine decades ago.)

On the other hand, some studies have shown a history of allergies as an adult, a mother eating fruits and vegetables during pregnancy, eating fruits and vegetables as a child, 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 now 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.

Environmental exposures are difficult to accurately measure, which can lead to inconsistencies across studies. A wide variety of environmental factors have been studied, such as vinyl chloride exposure; working in synthetic rubber manufacturing or petroleum refining/production; a history of head trauma, epilepsy, seizures or convulsions; cured food consumption (nitrites); viruses and common infections; cigarette smoking; alcohol consumption; residential power line exposure; exposure to air pollution; smoking when pregnant; second hand smoke exposure; agricultural worker exposures; industrial formaldehyde exposure; and the use of common drugs (for example, birth control pills, sleeping pills, headache medication, over-the-counter pain medication, antihistamines). However, additional, long term research on these factors is needed before definite conclusions can be formed.

**Genetic Factors**

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

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).

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.

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.

However, 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, and they may also interact with environmental factors as well. The Cancer Genome Atlas (TCGA) Project, a National Cancer Institute (NCI) and National Human Genome Research Institute (NHGRI) funded initiative, have 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 our 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.

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 oligodendroglialomas. 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.

**About Clusters of Brain Tumors**

We certainly understand community concerns when several individuals within a neighborhood are diagnosed with brain tumors. Scientists are interested in learning about these clusters of disease, but objectivity and the scientific process becomes very important in studying these groups.

The first piece of information scientists will want to learn is the “type” of brain tumors that seem to be occurring. Are these metastatic brain tumors—tumors that began as cancer elsewhere then move to the brain?

**Questions About Heredity**

“My family member has a brain tumor. Should I be tested too?” 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 relation 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 Cancer Information Service at 800-422-6237 can help you find a genetic counselor.

- **Young adult children can be taught their family history of brain tumor at the same time they learn their family history of other diseases, such as diabetes or high blood pressure.** They, too, can become good medical historians if they are equipped with honest and simple medical facts about their family.
Or, are these primary brain tumors—those that begin in the brain and tend to stay in the brain? Knowing the difference between these types of tumors is important to the scientists studying multiple occurrences of brain tumors.

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 tumors, 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.

From a patient or family member perspective, the first step in reporting a perceived cluster of brain tumors is a call to 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.

For More Information

To learn more about potential causes of brain tumors, you can perform a medical literature search on the Internet using PubMed, a medical literature search engine offered by the National Library of Medicine. PubMed can be found at www.nlm.nih.gov. This computer program searches medical journals (the journals scientists read) for articles containing the keywords and limits you specify. It has an easy “fill-in-the-blank” format and online help options.

Scientists are continually learning more about potential risk factors, and reporting results of their studies. To follow these reports, as well as other updates in the brain tumor community, the American Brain Tumor Association offers the ABTA News—an electronic monthly e-bulletin. Subscribe to this free update service at our web site: www.abta.org.
Sometimes a brain tumor is found by accident—it may be seen on a scan performed for a non-brain tumor purpose, such as a head injury. The tumor may not have been causing symptoms, or the symptoms were so minimal that you were not aware of the tumor’s presence until a scan was done.

Most commonly, a tumor makes its presence known with memory problems, changes in personality, changes in speech, a seizure, or one of the many other ways a brain tumor can interfere with the normal workings of the body.

Some of the same tests used to initially find your tumor will be used to monitor your progress. These tests will be used to see if the tumor disappeared, is shrinking, remains the same or has changed.

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 to make a diagnosis by taking your medical history. You are asked to describe your symptoms, how long you have had them, when they occur, if they seem to be brought on by something in particular, the order of their appearance, and if they seem to be getting worse. Following the question and answer phase of the process, your doctor will perform a basic neurological examination in the office.

Neurological Exam

A basic neurological examination includes the following:

- tests for eye movement by following a moving finger; pupil reaction, and eye reflex using a pen light
- vision tests and examination of the optic nerve
- hearing tests using a ticking watch or tuning fork
- reflex tests using a rubber hammer
- balance and coordination tests—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 tests using a sharp object and cotton ball or paint brush
- sense of smell tests with various odors
- facial muscle tests—smiling, grimacing
- tongue movement, gag reflex tests
- head movement tests
- mental status tests—i.e. stating the current time and date, naming the current President
- abstract thinking tests—i.e. defining the meaning of “a stitch in time saves nine”
- memory tests—i.e. 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. Those tests might include the following scans, xrays, or laboratory tests.

Scans

Scans take the place of conventional x-rays, which do not show tumors located behind the hard bones of the skull or spine. Different types of scanning, or imaging, devices are used to perform scans. The most commonly used imaging methods for diagnosis and follow-up are Magnetic Resonance Imaging (MRI) and Computerized Tomography (CT).

Both MRIs and CTs use computer graphics to create an image of the brain. An injection of a special contrast material (dye) to make abnormal tissue more obvious is usually given during the scan. The contrast materials concentrate in diseased tissues in greater quantity than in healthy tissues due to the leakiness of blood vessels in and around brain tumors. Contrast materials highlight abnormalities such as 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 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. 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 prior to contrast injection. If contrast is to be used, it is 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. 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).

MRI offers images with excellent anatomical detail which 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.

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

<table>
<thead>
<tr>
<th>MRI SCAN</th>
<th>MRI scan of an ependymoma. Scan courtesy of Dr. Regina Jakacki</th>
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<tr>
<td>TUMOR</td>
<td>This MRI scan shows an ependymoma.</td>
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<td>TUMOR</td>
<td>MRI scan of a glioblastoma multiforme. Scan courtesy of Dr. Jeffrey Bruce</td>
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<td>TUMOR</td>
<td>MRI scan of a single breast metastasis in the cerebellum. Scan courtesy of Dr. Deborah Heros</td>
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<tr>
<td>TUMOR</td>
<td>MRI scan of multiple brain metastases. Scan courtesy of Dr. Raymond Sawaya</td>
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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, however, 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 which feed the tumor, making surgery easier.

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

**PET (Positron Emission Tomography)**
PET 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 generation of 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 becomes combined with other types of scans, as mentioned above.

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). 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. However, 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 COMPUTORIZED TOMOGRAPHY)**

SPECT 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)**

MEG scans measure 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. However, as sophisticated as this sounds, MEG scans do not “read” or record your thoughts.

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.

MEG is 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, however, as government support for the development of this technique increases, community access may also increase.

**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. X-rays 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 miniscule 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 which 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.

<table>
<thead>
<tr>
<th>Hormone</th>
<th>Responsible for...</th>
<th>Normal Blood Levels in Adults</th>
</tr>
</thead>
<tbody>
<tr>
<td>ACTH (adrenocorticotropic hormone)</td>
<td>Controls the production of cortisol — a natural steroid needed to control blood pressure, sugar and salt levels</td>
<td>9 to 52 pg/ml</td>
</tr>
<tr>
<td>GH (growth hormone)</td>
<td>Controls bone growth; height; body proportion in the extremities and jaw</td>
<td>0 to 3 ng/ml</td>
</tr>
<tr>
<td>PRL (prolactin)</td>
<td>Controls milk production in women, impacts sex drive and sperm counts in men</td>
<td>Males and non-pregnant women: 0 to 20 ng/ml In pregnancy: 10 to 300 ng/ml</td>
</tr>
<tr>
<td>TSH (thyroid stimulating hormone)</td>
<td>Controls thyroid functions such as metabolism, heart rate and appetite</td>
<td>0.2 to 4.7 mU/l</td>
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</table>
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. 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 every 3 month, 6 month, or perhaps yearly MRIs. Even those whose tumors were treated ten ++ years ago should still be followed by a physician who knows about the tumor. Very late recurrences can happen; knowing your history can help a doctor 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.

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 to be 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. Tumor tissue is removed from the core of the needle.

Stereotaxic 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.
Types of Brain and Spinal Cord Tumors

This chapter contains, in alphabetical order, information about the more common brain and spinal cord 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.

In addition, we offer more detailed information about specific tumors and treatments at our web site—www.abta.org.

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

Also called Neurilemmoma, Vestibular Schwannoma or Neurinoma

The acoustic neuroma is a benign tumor of the nerve of hearing (the 8th cranial nerve). It is located in the angle between the cerebellum and the pons, in the posterior fossa (the back of the skull). This tumor usually grows very slowly.

Acoustic neuromas typically occur in adults, particularly in their middle years. Females are twice as likely to have this tumor as males. Acoustic neuromas account for about 8% of all primary brain tumors.

Common symptoms of this tumor are one-sided hearing loss, and buzzing or ringing in the ear. Dizziness may also occur but is less common. If the tumor also affects the facial nerve (the 7th cranial nerve) located next to the 8th nerve, facial paralysis may occur. Other symptoms include difficulty in swallowing, impaired eye movement, taste disturbances, and unsteadiness.

Total removal using microsurgical techniques is often possible. Stereotactic radiosurgery might be used as an alternate to surgery for some patients.

Tumors on both sides (bilateral) are rare, and tend to be familial. They are almost always associated with neurofibromatosis 2, a hereditary condition. The malignant form of this tumor, malignant peripheral nerve sheath tumor (MPNST), is extremely rare.

For more information on acoustic neuromas, contact:

Acoustic Neuroma Association
600 Peachtree Parkway, Suite 108
Cummings, Georgia 30041
Phone: 770-205-8211
Website: www.anausa.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. The WHO system grades astrocytomas on a scale from I to IV. Grade I tumors include pilocytic astrocytomas, which are usually localized tumors and are often cured with 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 the various grades of these tumors. The list begins with grade I tumors and progresses through grade IV astrocytomas.

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 (ie, an “optic glioma”), the optic chiasm near the hypothalamus, thalamus, basal ganglia, cerebral hemispheres, and the cerebellum (ie, 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 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 re-growth. 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 (called necrosis). These rare tumors require more aggressive treatment than a benign pilocytic astrocytoma.

SUBEPENDYMYAL GIANT CELL ASTROCYTOMA

Subependymal giant cell astrocytomas are ventricular tumors associated with tuberous sclerosis. *Please see the section on tuberous sclerosis for additional information.*

**Diffuse Astrocytoma**

Also called Astrocytoma, Low Grade or Astrocytoma Grade II (types: Fibrillary, Gemistocytic, Protoplasmic Astrocytoma)

These low grade astrocytomas tend to be infiltrating tumors, capable of growing into surrounding tissue, but tumors which grow relatively slowly. Astrocytomas are grouped by the appearance and behavior of the cells for which they are named. For example, the nuclei of fibrillary astrocytoma cells are cigar-shaped. That type of astrocytoma tends to contain microcysts and mucous-like fluid. The nuclei of protoplasmic astrocytoma are round or oval in shape. Those tumors also tend to contain microcysts and mucous-like fluid. Gemistocytic astrocytomas have plump, glassy, angular shaped cells. (Until recently there had been some disagreement as to the grade of the gemistocytic astrocytoma. Some pathologists considered this to be a grade III tumor since the gemistocytic astrocytoma, in particular, tends to recur as an anaplastic astrocytoma. The WHO classification system published in 2000 defines these as grade II tumors.)

Regardless of the cellular appearance of these grade II astrocytomas, surgical removal may be suggested for accessible tumors (tumors that may be removed without causing undue neurological damage). If “total” surgical removal can be achieved, periodic follow-up with MRI or CT scans might be the only additional care required. It is important to realize that “total” removal generally means removal of all of the tumor visible to the neurosurgeon’s eye. Microscopic cells, too small to see, can remain behind or may have spread to nearby tissue. Those cells may later begin to re-grow and in some cases these tumors can transform into grade III or IV tumors. However, even with incomplete removal these tumors tend to grow very slowly, and it may be several years before there are imaging changes or symptoms from tumor re-growth.

In adults and older children, radiation therapy may be suggested in addition to surgery, or radiation may be used to treat a low grade astrocytoma which is unable to be removed. The role of chemotherapy in treating these tumors is under investigation. Further treatment might be recommended only if the tumor recurs. Children younger than three might receive chemotherapy so that radiation can be delayed.

These low grade tumors may recur as a higher grade tumor; thus, periodic follow-up and attention to the return of symptoms is important. Treatment of the recurrent tumor would be based on the tumor’s grade at the time of re-growth.
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 looked 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 beam radiation 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; there are also many new drugs being tested in clinical studies. In addition, 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.

We offer additional information about anaplastic astrocytoma and glioblastoma (a grade IV astrocytoma) at our web site—www.abta.org—and as a print booklet. Please call our office if you would like a copy of that information.

ASTROCYTOMA GRADE IV
Also called Glioblastoma, previously named “Glioblastoma Multiforme”

“Grade IV astrocytoma,” “glioblastoma,” and “GBM” are all names for the same tumor. Glioblastomas represent about 17% 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 technically can be found anywhere in the brain or spinal cord. Because the glioblastoma is capable of very rapid growth, the first symptoms are usually due to increased pressure in the brain. Headaches, seizures, memory loss, and changes in behavior are the most common presenting symptoms.

A glioblastoma commonly contains 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 generally made when several criteria are observed upon microscopic examination: the tissue sample contains highly malignant astrocytic cells, an actively growing blood supply, and a high
percent of tumor cells reproducing at any given time. 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. Dead (necrotic) cells may also be seen, especially toward the center of the tumor.

In recent years, advanced biotechnology has allowed glioblastomas to be sub-divided into two groups: primary and secondary glioblastoma. Primary, or de novo, glioblastoma are grade IV tumors from the onset. These are most likely the very aggressive, most common form of glioblastoma. Secondary glioblastoma are those that originate as a lower grade tumor (grade II or III) and then evolve into a grade IV tumor. 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 subcategories of glioblastoma. That information may also soon lead to other sub-groupings of glioblastoma, and therapies specific to those biological differences between tumors.

Lack of exactly the same cells throughout 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. This is often referred to as “tumor cell heterogeneity.”

The first step in treating a glioblastoma is surgery to remove as much tumor as possible. Radiation therapy, given with a drug called temozolomide, almost always follows surgery or biopsy. There are several different forms of radiation therapy —your doctor will suggest the type of radiation best for your tumor.

The most commonly used chemotherapy drug in adults is currently temozolomide, administered concurrently with radiation and then given for 6 – 12 months after completion. However, other drugs are also being tested. Many of the these 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 prefer 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 re-growth 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 interfere with, or block, cell signaling pathways which control cell reproduction, invasion into the surrounding brain tissue and abnormal development of blood vessels. The ability to identify these biologic differences, and to create drugs that target the differences, is the field of medicine referred to as “personalized” or “individualized” medicine. While this is an exciting area of science, these drugs are just beginning development and testing.

Immunotherapy—the use of vaccines or immunizations—is another area of great 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 the programs are open to those with a glioblastoma. Some of these studies 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 diptheria 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 targets the radiation to the tumor cells.

These new therapies are offered in organized research studies called clinical trials. Many clinical trials are available for glioblastoma—both as initial treatment and as treatment for a
Many of those clinical trials can be found through the National Cancer Institute’s Cancer Information Service at 800-422-6237. There are also many online services providing clinical trial information; please call our office at 800-886-2282 for those resources.

Additional information about glioblastoma is offered at our web site—www.abta.org.

**Atypical Teratoid Rhabdoid Tumor (ATRT)**

This rare, high grade tumor occurs most commonly in children younger than three years of age, but on rare occasions are diagnosed in adults. ATRTs are generally found in the cerebellum—the lower, back part of the brain which controls balance. These tumors tend to be aggressive and frequently metastasize (spread) through the central nervous system. Previously thought to be PNETs (primitive neuroectodermal tumors) or medulloblastoma tumors, ATRTs were found to have their own histologic and cytologic features which now help a pathologist specifically define this tumor.

Treatment generally involves removal of the tumor followed by chemotherapy. Radiation therapy may be considered depending on the age of the child, and whether the tumor has recurred. Clinical trials are underway to help develop chemotherapy drugs effective against this tumor—those clinical trials can be found by calling the Cancer Information Service at 800-422-6237.

**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. However, in these situations, the diagnosis can usually be based on the MRI scan features.

Most of these tumors are classified by their location:

- 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 the brainstem)

The majority of brain stem tumors occur in the pons, are diffusely infiltrating, and therefore are not able to be surgically removed. A few of these tumors are localized, and may be reachable for resection. These tumors tend to be very slow growing, not in the pons, and are exophytic—on the outer edges of the brain stem.

The symptoms of a brain stem glioma depend on the location of the tumor. The most common symptoms are related to eye movement abnormalities and 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, the grade and the symptoms. Surgery may be warranted if a tumor appears circumscribed (contained) or exophytic (on the outside of the
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 available.

Chondroma

This rare, benign tumor tends to arise at the base of the skull, especially in the area near the pituitary gland. A chondroma is generally very slow growing and might be present for a long time before causing any symptoms.

These tumors are composed of cartilage-like cells and are usually attached to the dura mater, the outermost layer of the meninges. A chondroma can grow to a large size, and may occur as a single or as multiple tumors. The malignant form of this tumor is called a chondrosarcoma (see the chondrosarcoma section which follows).

Because it is usually accessible with well-defined margins, surgery might be the only treatment required for a chondroma.

Chondrosarcoma

This very rare tumor arises from bone and is composed of cartilage. It is the malignant variant of the benign chondroma (described above) and tends to spread locally, staying within the same general area. This tumor is generally slow growing and rarely metastasizes, or spreads, to areas farther away. It is most commonly found in the sphenoid bone—the bony ridge running along the back of the eyes—or near the clivus, a bony area at the base of the skull. The chondrosarcoma is more common in adult males.

Standard treatment is surgical removal which might be followed with radiation therapy. Those with a chondrosarcoma may also be eligible for treatment in a clinical trial—an organized research study. Those studies can be found through the Cancer Information Service at 800-422-6237.

Chordoma

The chordoma occurs at the base of the skull in about one-third of patients, or at the end of the spine. It is a benign, slow growing tumor. When found in the spine these tend to be extradural tumors, meaning that they are located on the outside of the spinal cord. However, a chordoma may invade the nearby bone, compressing parts of the brain in the area. It is not unusual for a chordoma to push into the brainstem or grow into the sinuses. Distant spread is rare.

This is an uncommon tumor representing only 0.2% of all primary CNS tumors. Although it is found in people of all ages, chordomas are most frequent in younger and middle-aged adults. The most common symptoms are double vision and headache.

The chordoma is visible on MRI scans, but a biopsy is necessary to determine an exact diagnosis. The skull base location can be very difficult to access, making complete removal of tumors in this area a challenge. A combination of surgery followed by radiation is the standard treatment for tumors located in the skull base. Stereotactic radiosurgery and stereotactic radiotherapy have shown promise, as has the combination of aggressive surgery followed by combined proton-photon beam therapy. Complete surgical resection of visible tumor might be possible for the spinal chordoma.
Choroid Plexus Tumors, Choroid Plexus Papilloma, Choroid Plexus Carcinoma

This group of tumors, which occur primarily in children, comprise about 10 – 20% of all brain tumors diagnosed in children during their first year of life, and 2 – 4% of those tumors found in children under age 15. These tumors arise from brain tissue called the “choroid plexus,” which lines the ventricles of the brain and produce cerebrospinal fluid. Lateral and third ventricle tumors tend to be found in very young children. Fourth ventricle and cerebellopontine angle tumors (the angle between the cerebrum and the pons of the brain stem) generally occur in those ages 20–35. The diagrams in the Parts of the Brain section of this book will help you visualize these locations.

Like other brain tumors, choroid plexus tumors are also “graded.” Grade is based on how unusual their cells appear when viewed under a microscope. Choroid plexus papillomas are grade I tumors. Atypical choroid plexus papillomas are considered grade II, and choroid plexus carcinomas are generally grade III tumors.

Choroid plexus papilloma is a rare, benign tumor which grows slowly within the ventricles. It may eventually block the flow of cerebrospinal fluid, causing hydrocephalus and increased intracranial pressure. Headache and other symptoms of increased pressure are common. Standard treatment is surgery to remove as much tumor as safely possible. During tumor removal, a surgical neuroendoscope may be used by the neurosurgeon to reach tumor in the passageways of the ventricles. Surgery may be the only treatment required if the tumor is completely removed. Tumor removal relieves the hydrocephalus about half of the time; a shunt may be required for the other patients. The role of radiation or chemotherapy is still being defined, but might be recommended for inaccessible or partially resected tumors.

Choroid plexus carcinoma, which occurs primarily in children, is the malignant form of the choroid plexus papilloma. It comprises about 10% of all choroid plexus tumors and typically occurs in one of the lateral ventricles. (The choroid plexus carcinoma is sometimes called an anaplastic choroid plexus papilloma.) These tumors commonly invade, or grow into, nearby tissue and spread widely via the cerebrospinal fluid. Hydrocephalus—a collection of cerebrospinal fluid within the brain—is often present. Treatment often includes surgery, chemo-therapy and radiation therapy. A second surgery might be recommended for recurrent tumors, followed by some form of radiation and/or chemotherapy.

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 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 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 intracavitary use of radioactive phosphorous. In children younger than 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.

Cysts

Just like a cyst elsewhere in your body, a cyst in the brain is a tumor-like sphere filled with fluid, similar to a balloon filled with water. Cysts may contain fluid, blood, tissue, or tumor cells. There are specific types of cysts; they are named for the type of tissue from which they arose and for their contents. The most common cysts found in the brain are arachnoid, colloid, dermoid, and epidermoid cysts. Each is described below.

**Arachnoid Cyst**

An arachnoid cyst (sometimes called a leptomeningeal cyst) is an enlarged, fluid-filled area of the subarachnoid space—the space between the arachnoid and pia mater layers of the meninges which form a membrane-like covering around the brain and spinal cord. Arachnoid cysts occur in both adults and children. Their most common locations are in the area of the Sylvian fissure, the cerebellopontine angle, the cisterna magna or the suprasellar region of the brain. Treatment may be “watchful waiting,” or the cyst may require surgery. The goal of surgery, if needed, is to attempt to remove the entire cyst, including its outermost lining. If that is not feasible, the neurosurgeon open the cyst wall to drain the contents. If the cyst is blocking the flow of cerebrospinal fluid, a shunt may be used to help divert the fluid to other areas of the body.

**Colloid Cyst**

Although scientists are not sure of the definitive cause of colloid cysts, most agree that these cysts begin during embryonic development of the central nervous system. These cysts tend to contain a thick, gel-like substance called colloid. The roof of the third ventricle is the most frequent location of this cyst. Malignant forms are unknown. Colloid cysts almost always occur in adults. They are typically attached to the roof of the third ventricle and the choroid plexus. This location may block the flow of fluid through the foramen of Monro, one of the small tunnels through which cerebrospinal fluid exits the ventricles, causing increased intracranial pressure. Headache is the most common symptom.

Various surgical approaches, stereotactic directed cyst drainage or shunting are some of the treatment options for the colloid cyst. Removing this cyst without causing undue damage can be challenging because of its location on/near the third ventricle, and the “best” treatment is dependent on individual patient’s anatomy and the configuration of their cyst.

**Dermoid Cyst**

Dermoid cysts likely form during the early weeks of fetal development even though the symptoms may not be noticed for years after birth. As an embryo is developing, the neural tube—the cells which will eventually form the brain and spine—begins to separate from the cells which will become the skin and bones of the face, nose, and vertebrae. A dermoid cyst results when cells that normally belong to the face are diverted to the brain or the spinal cord. That’s why the inside of a dermoid cyst often contains hair follicles, bits of cartilage, or sebaceous glands which produce skin oils and fats. On rare occasions, a dermoid cyst may spontaneously open, releasing these oils into the brain or spinal cord.

Dermoid cysts are relatively rare masses to be found in the brain—epidermoid cysts are far more common. However, when a dermoid cyst is found in the brain it is usually a benign mass. These cysts are usually located in the posterior fossa (the lower back portion of the brain) or the adjacent meninges (the thin membranes which form the covering of the brain and spinal cord). Dermoid cysts in the brain tend to occur in children under 10 years old. The lower end of the spine is the more
common location in older children and young adults. The cavity of the fourth ventricle and the base of the brain under the surface of the frontal lobes are also common sites.

The standard treatment for a dermoid cyst is surgical removal. If the cyst is unable to be completely removed, it will likely regrow. That growth may be very slow, and it may be years before symptoms would again return.

**EPIDERMOID CYST**

*Also called Epidermoid Tumor*

Epidermoid cysts, also referred to as epidermoid tumors, likely form during the early weeks of fetal development even though the symptoms may not be noticed for several decades into life. As an embryo is developing, the neural tube—the cells which will eventually form the brain and spine—begins to separate from the cells which will become the skin and bones of the face, nose, and vertebrae. An epidermoid cyst results when cells that normally belong to the face and bones are diverted to the brain. That’s why the inside of an epidermoid tumor often contains remnants of skin cells or tiny pieces of cartilage. On rare occasions, an epidermoid cyst may spontaneously open, releasing these contents into the brain or spinal cord.

Epidermoid cysts occur more frequently in the brain than in the spine, and are a fairly common type of cyst to be found in the brain. They are usually benign, and are most commonly found in middle-aged adults. These cysts tend to be located near the cerebellopontine angle (the area where the top part of the brain meets the brain stem) and near the pituitary gland. Standard treatment is surgical removal. If the cyst is unable to be completely removed, it can re-grow. That growth may be very slow, and it may be years before symptoms would again return.

**Dysembryoplastic Neuroepithelial Tumor (DNT)**

DNTs are slow growing, benign, grade I tumors. Traditionally seen as similar in appearance and behavior to an oligodendroglioma, laboratory testing allows pathologists to separate this tumor from other similar appearing tumors. These tumors tend to contain a mix of neurons (nerve cells) and glial (supportive) cells suspended in a mucous-like substance.

Although they occur in both adults and children, this tumor tends to be found in people ages twenty to thirty. It is not unusual for the diagnosis to be preceded by a long history of uncontrollable seizures of the partial complex type. DNTs have been occasionally associated with a history of neurofibromatosis type I.

The DNET is most commonly located in a temporal or frontal lobe of the brain. Surgery alone often provides long term control for this tumor, even when the tumor is unable to be completely removed.

**Ependymoma**

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 5–6% of all gliomas. However, they are the most common brain tumor in children, and 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*, *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 most common of the ependymal tumors, and are considered grade 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 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.

We offer additional publications specifically about ependymoma and brain tumors in children. Please call our office at 800-886-2282 if you would like that information.

For more information on ependymomas, contact:

CERN Foundation (Collaborative Ependymoma Research Network)
6450 Poe Avenue, Suite 201
Dayton, OH 45414
Phone: 937-264-2574
Website: www.cern-foundation.org

Gangliocytoma and Ganglioglioma

These rare, benign tumors arise from ganglion-type cells, which are groups of nerve cells. Gangliocytomas (sometimes called ganglioneuromas) are tumors of mature ganglion cells. Gangliogliomas are tumors of both mature nerve and supportive cells (glia).}

Gangliocytomas and gangliogliomas most frequently occur in children and young adults. They represent less than 1% of all primary brain tumors, about 4% of all pediatric brain tumors, and tend to occur more often in females.

These tumors are most commonly located in the temporal lobe of the cerebral hemispheres and the third ventricle, although they might also occur in the spine. These tumors are small, slow growing, and have distinct margins. Metastasis and malignancy are very rare.

Cyst formation and calcification can be present. Seizures are the most common symptom.

The standard treatment for gangliocytoma and ganglioglioma is surgery.

Germ Cell Tumors

See also pineal region tumors, germinoma, and teratoma

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 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, although a biopsy to establish an exact diagnosis is not uncommon and some very experienced surgeons have had success removing certain pineal region tumors. Surgery may be required to treat hydrocephalus caused by a blockage, by the tumor, of the cerebrospinal fluid pathways.

**Germinoma**

The germinoma is the most common type of germ cell tumor in the brain. It typically occurs in the pineal or suprasellar region of the brain. Because it tends to spread via the cerebrospinal fluid, 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.

The germinoma is the most common tumor of the pineal region representing about 30% of those tumors. Its occurrence is more usual in teen-aged children, and in males more often than females.

Tumors in the pineal region typically cause symptoms indicating increased intracranial pressure. Headache due to obstructed cerebrospinal fluid flow is the most common symptom. If the tumor is in the suprasellar location, symptoms include diabetes insipidus, vision changes, signs of hormonal dysfunction such as fatigue, poor appetite, delayed or absent puberty, and changes in the menstrual cycle.

Surgery for germinoma depends on its accessibility and position relative to critical brain structures; most often only an open biopsy is performed as surgical risk should be minimized due to the responsiveness of this type of tumor to medical treatment. The germinoma is very responsive to radiation and this can be an effective treatment for some patients. Chemotherapy might be the treatment of choice for some newly diagnosed tumors. Chemotherapy can also be useful for recurrent tumors.

**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 17% 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 subdivided 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 tumors which 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 these 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 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 create drugs that target the differences, is the field of medicine is referred to as “personalized” or “individualized” medicine. While this is an exciting area of science, these drugs are just beginning development and testing.

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 diptheria 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. Many clinical trials are available for glioblastoma—both as initial treatment and as treatment for a recurrent tumor. Those clinical trials can be found through the National Cancer Institute’s Cancer Information Service at 800-422-6237. There are also many online services providing clinical trial information; please call our office at 800-886-2282 for those resources.

We can provide additional information about glioblastoma, new treatment options, support resources, and family/caregiver support services. Please contact us to access these, and many other, ABTA services.

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 oligodendrogliomas, 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.

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.

Glioma, Mixed

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

Standard treatment for a mixed glioma is similar to that for astrocytoma and oligodendroglioma of the same grade. The treatment plan may includes surgery followed by radiation therapy, particularly if the tumor is high-grade (grade III or IV).
Glioma, Optic

These tumors may involve any part of the optic pathway including the optic nerve right behind the eyeball, the optic chiasm where both optic nerves come together, the optic tracts located close to the brain stem or the optic radiations within the brain. Optic gliomas also have the potential to spread along these pathways. Most of these tumors occur in children under the age of 10. The grade I pilocytic astrocytoma and grade II fibrillary astrocytoma are the most common tumors affecting these structures. Higher-grade tumors may also arise in this location.

Twenty percent of children with neurofibromatosis 1 (NF-1), a genetic disorder affecting the skin and nervous system, will develop an optic glioma. These gliomas are typically grade I, pilocytic astrocytomas. Children with optic gliomas are usually screened for neurofibromatosis (NF) for this reason. Adults with NF-1 generally do not develop optic gliomas.

These tumors may be quiet, causing few or no symptoms. Their placement along the nerves of seeing, however, can cause loss of vision (in one eye or partial vision loss in both eyes depending on the location of the tumor) or strabismus (“crossed eyes”). Hormonal disturbance might also occur causing developmental delay, early puberty and other symptoms.

Careful observation may be an option for patients with stable or slow growing tumors. Treatment other than monitoring is based on symptoms or changes seen on the MRI scan. Surgery might be recommended for a growing tumor which involves only the optic nerve. Radiation therapy might be used for a tumor of the chiasm or other pathways. Local radiation therapy and chemotherapy with radiation therapy are used for recurrent tumors. Clinical trials are available for both primary and recurrent tumors.

Gliomatosis Cerebri

This condition is an uncommon primary brain tumor characterized by a diffuse, or broad, spread of glial tumor cells in the brain. This tumor is distinct from other gliomas because of its scattered and widespread nature, typically involving two or more lobes of the brain. It could be considered a “widespread low-grade glioma” because it lacks the malignant features (such as abnormal blood vessels’ growth and dead tissue) seen with high-grade tumors. The diffuse nature of gliomatosis causes enlargement of any part of the brain it involves including the cerebral hemispheres, or less often, the cerebellum or the brain stem. Symptoms are often nonspecific and can include personality and behavioral changes, memory disturbance, increased intracranial pressure with headache and sometimes seizures. Treatment is less well-defined given the rarity of this tumor. Surgical resection is generally not attempted due to the diffuse nature of the tumor; the diagnosis might be based on biopsy only. Radiation therapy and chemotherapy may be considered.

Glomus Jugulare

See also Paraganglioma

Glomus jugulare tumors are also called paragangliomas. They originate from chemical sensors called chemoreceptors located in the lining of a large vein in the neck called...
the jugular vein. These tumors are rare, slow growing and are usually benign, but can spread to the bone close to the inner and middle ear. This tumor is most often found in people about 50 years old. Most patients have symptoms related to the inner ear including hearing loss, abnormal noise in the ear, dizziness, ear pain or bleeding in the external ear canal. Other nerves located close to the jugular vein can be affected and result in facial weakness, hoarseness or swallowing difficulties.

The initial diagnosis of this type of tumor may be made with an MRI scan which shows a tumor either at the jugular vein opening at the base of the skull or higher up, closer to the inner ear canal, cerebellum and brainstem. Cerebral angiography using a dye injected in brain vessels can be helpful in making a diagnosis because this tumor often has a large blood supply. If the tumor “runs in the family,” it is not unusual to find multiple tumors in the same person.

Because of the rarity of this tumor the most effective treatment is not well-defined. Surgery appears to benefit young patients who have a large tumor causing symptoms. The surgical team often consists of a neurosurgeon and a “head and neck” surgeon. (“Head and neck” is not the same as the “brain.” These are separate areas of medicine.) Standard radiation or radiosurgery also appears effective in stabilizing these tumors. Radiosurgery may be used in patients with small tumors or when residual tumor remains after surgery.

**Hemangioblastoma**

Hemangioblastoma is a benign and slow-growing tumor arising from cells in the blood vessel lining. These cystic tumors tend to have clearly indicated borders and do not infiltrate the surrounding normal tissue. Single or multiple tumors may be present. They are most common in the lowest part of the brain—the posterior fossa, which contains the cerebellum and the brainstem—but can occur in the cerebral hemispheres, the spinal cord or even the retina.

Hemangioblastomas represents about 2% of all primary brain tumors. About 10% of patients with hemangioblastoma have Von Hippel-Lindau disease, an inherited condition that predisposes to this tumor as well as tumors of the liver, pancreas and kidneys. The tumor can occur at any age but most commonly found in people about 40 years of age.

This tumor commonly causes blockage of the cerebrospinal fluid circulation resulting in hydrocephalus (enlarged fluid cavities of the brain) and increased intracranial pressure. It may also compress the cerebellum which is involved in balance and limbs coordination. The most common symptoms include headache, nausea and vomiting, gait disturbances, and poor coordination of the limbs.

MRI scans can be used to image the hemangioblastoma. Angiography sometimes is done before surgery to confirm the diagnosis and provide information about the tumor’s blood supply.

Surgery is the standard treatment for hemangioblastoma. Incompletely removed tumors or tumors attached to the brain stem might be treated with a form of focused radiation such as stereotactic radiosurgery. Drugs affecting the growth of the blood vessels which feed the tumor (a process called angiogenesis) are being studied for this disease.

For more information about Von Hippel-Lindau disease, contact:

VHL Family Alliance
171 Clinton Road
Brookline, Massachusetts 02445
Phone: 800-767-4845
Website: [www.vhl.org](http://www.vhl.org)

**Hemangiopericytoma**

This is a rare grade II or III tumor, different in origin from meningiomas although arising in the same location—the lining of the brain called meninges. These tumors appear to originate in the cells surrounding the blood vessels in the meninges, and this explains the large blood supply within these tumors. They do not invade the brain but have a greater potential than meningioma to recur locally or even spread elsewhere in the body (bone, lung and liver) although the latter does not occur until late in the disease. Hemangiopericytomas affect younger people than meningiomas, and
people usually have symptoms for less than a year before diagnosis.

**Lipoma**

Lipomas are rare, benign tumors composed of fatty tissue. The most common location is in the region of the corpus callosum, but they also occur in other areas in the brain usually close to the midline (the middle part of the brain where the two hemispheres of the cerebral lobes meet). A lipoma may cause no symptoms and is often diagnosed coincidentally when scans are performed for other medical reasons. Some lipomas are associated with other congenital abnormalities of the nervous system. It is diagnosed by either CT or MRI scanning. Conservative treatment is usually recommended since these tumors rarely cause symptoms. Surgery may be suggested in some circumstances.

**Lymphoma**

*Also called CNS Lymphoma, Primary Malignant Lymphoma or Primary CNS Lymphoma (PCNSL)*

This disease affects people with healthy immune systems as well as those whose immune systems are not functioning properly—such as organ transplant recipients or people who are HIV positive. CNS lymphoma most commonly originates from B lymphocytes and is classified as non-Hodgkin’s (meaning it is different from Hodgkin’s disease). The incidence of CNS lymphoma has been increasing over the past 20 years; it now represents between 0.5% and 2% of all primary brain tumors. The increase is thought to be related to the AIDS epidemic, but not all people diagnosed with lymphoma are HIV positive.

Lymphoma occurs most often in the cerebral hemisphere but may also involve the cerebrospinal fluid, the eyes or the spinal cord. In addition, a few people may have evidence of lymphoma elsewhere in the body. It is not unusual for this tumor to be found in multiple places in the cerebral hemisphere as it does have the potential to spread throughout the central nervous system.

The most common symptoms of CNS lymphoma include personality and behavioral changes, confusion, symptoms associated with increased intracranial pressure (headache, nausea, vomiting, drowsiness), weakness on one side of the body and seizures. Problems with eyesight, such as blurred vision, floaters or double vision may also occur.

MRI scans are used to diagnose the presence of this tumor, but a biopsy is required for confirmation since other types of tumor, infections or inflammatory disorders may look the same on the scan. Steroids should be held until after the biopsy, if at all possible. A spinal tap to screen for tumor cells in the cerebrospinal fluid (CSF) might be performed as long as there is no indication of increased intracranial pressure. However, a negative CSF study does not rule out lymphoma as it is positive in only about one-third of cases.

If the biopsy is positive for lymphoma, examination of other parts of the body, including the eyes, will be done to determine if the tumor has spread. This testing may include a CT scan of the chest, abdomen and pelvis, blood tests, and a spine MRI if warranted. Sometimes, a whole body PET scan and/or bone marrow biopsy may be done. Because of an increased incidence of testicular cancer and lymphoma, a testicular ultrasound may be suggested for those under 60 years of age.

After the diagnosis of lymphoma is confirmed, steroids are used to control brain swelling; this may result in the immediate disappearance of the tumor on a subsequent scan. Chemotherapy and radiation, or chemotherapy alone may then be used as the primary treatment. Lymphomas are not usually treated with surgery as they tend to occur deep within the brain in which case the risk of neurological complications is high.

**Medulloblastoma (MDL)**

Medulloblastoma represents about 13% brain tumors in children under the age of 14. In addition, about 20% of these tumors occur in adults. Medulloblastoma is 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, 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 headache, nausea, vomiting and drowsiness, gait unbalance and poor coordination of the limbs. Unusual eye movements may also occur.

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 this would include an MRI of the spine and a cerebrospinal fluid analysis. For older children, adults without evidence of spread 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 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. To learn more about clinical trials for either pediatric or adult medulloblastomas, contact the Cancer Information Service at 800-422-6237.

We offer additional publications specifically about medulloblastoma and brain tumors in children. Please call our office at 800-886-2282 if you would like that information.

Meningioma

These tumors arise from the “arachnoid mater” —one of the layers of the meninges (the lining of the brain). Meningiomas represent about 34% 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 (“parasaggital meningiomas”) or over (“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” (see section on NF) 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 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, or 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 rapidly 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.

We offer additional information about meningioma. Please call our office at 800-886-2282 if you would like that material.

**Metastatic Brain Tumor**

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

We offer a publication specifically about metastatic brain tumors. Please call our office at 800-886-2282 for a copy of that resource.

**Neuroblastoma, Cerebral**

Neuroblastoma most commonly occurs outside the central nervous system, in the abdomen and chest. It is part of the family of tumor called primitive neuroectodermal tumors or “PNET.”

When found in the brain, these are usually malignant, rapidly growing tumors. They tend to occur in the cerebral hemisphere, and may cause increased pressure within the brain, seizures or weakness on one side. Like the PNET, cerebral neuroblastoma has the potential to spread throughout the central nervous system via the cerebrospinal fluid. The assessment and treatment are identical to other PNET and outlined in that section.

**Neurocytoma, Central**

This rare, grade II tumor typically occurs in the fluid cavities of the brain, called the lateral ventricles, in the region of the foramen of Monro (the area where the left and the right lateral ventricles come together). It occasionally extends into the third ventricle as well. The central neurocytoma contains mature cells similar to normal neurons—the “thinking cells” of the brain—although their exact cell of origin is unknown. Central neurocytoma is most common in young adult males. These tumors usually block the cerebrospinal fluid circulation causing hydrocephalus (dilatation of the brain fluid cavities). Resulting symptoms are those associated with increased intracranial pressure, such as headache, nausea, vomiting, drowsiness. Seizures may also occur.

Standard treatment is surgery, which is often successful; however, excessive bleeding can limit the extent of tumor removal. The role of radiation therapy is unclear. However, if the tumor has more aggressive features or if the tumor recurs, radiation therapy may be recommended.

**Neurofibromatosis (NF)**

The term neurofibromatosis refers to two different genetic diseases characterized by skin abnormalities and nervous system tumors. Neurofibromatosis type 1, also called NF-1 or Von Recklinghausen’s disease, is the more common of the two disorders. It causes tumors of the nerves, called neurofibromas, throughout the body and often visible underneath the skin. It also causes skin discolorations called “café-au-lait” spots as well as freckles in the arm pits and groin regions. Other nervous system tumors can be associated with NF-1; these occur in approximately 10% of patients and include optic pathway gliomas (usually pilocytic astrocytoma), cerebral hemisphere, posterior fossa (brain stem and cerebellum), and low-grade astrocytomas in the spinal cord.

NF-2-associated nervous system tumors may include tumors of the hearing nerve (acoustic neuromas or vestibular schwannoma) typically
on both sides, meningiomas, schwannomas of the spinal root of the nerves, and ependymomas (spinal cord or brain).

The predisposition to tumor formation in both disorders is related to genetic abnormalities which interfere with a protein that normally regulates and prevents tumor formation.

Extensive information about neurofibromatosis is available from:
The National Neurofibromatosis Foundation, Inc.
95 Pine Street, 16th Floor
New York, New York 10005
Phone: 800-323-7938
Website: www.nf.org
or
Neurofibromatosis, Inc.
9320 Annapolis Road, Suite 300
Lanham, Maryland 20706
Phone: 800-942-6825
Website: www.nf inc.org

**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.

Oligodendrogliomas can be low-grade (grade II) or high-grade (grade III also called anaplastic). Sometimes oligodendrogliomas 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.

Oligodendrogliomas 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 these tumors being found in the frontal lobe. Seizure is 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 oligodendroglioma that appear on the MRI scan after surgery to have been completely resected, close observation with follow-up MRIs may be recommended. If some of the tumor remains after surgery (this is called “residual” tumor), radiation therapy appears to be indicated although the best timing—immediately or at tumor progression—is being determined in clinical trials. Recurrent low-grade oligodendrogliomas can be treated with surgery, radiation therapy (if not given initially), or chemotherapy.

For anaplastic oligodendroglioma, a combination of radiation therapy and chemotherapy such as PCV (procarbazine, CCNU and vincristine) or temozolomide is indicated. Recurrent anaplastic oligodendroglioma may be treated with surgery and/or chemotherapy. Genetic analyses of oligodendroglioma have shown that combined loss of the short arm of chromosome 1 and the long arm of chromosome 19 (called “1p 19q loss”) is associated with improved outcome. Clinical trials are available for newly diagnosed and recurrent, low-grade or high-grade oligodendrogliomas. Many of these trials take into account the genetic features of the tumor, thereby highlighting the importance of obtaining tumor tissue for analysis.

We offer additional information about oligodendroglioma. Please call our office if you would like that material.
Pineal Tumors

The pineal gland is located at the rear of the third ventricle—one of the fluid cavities of the brain. Pineal region tumors represent less than 1% of all primary brain tumors; however, 3% to 8% of childhood brain tumors occur in this area. Several tumor types may occur in this area:

- Germ cell tumors including germinoma (tumor similar to other occurring in testes or ovaries), and non-germinoma (including several such as teratoma, endodermal sinus tumor, embryonal cell tumor, choriocarcinoma and mixed tumors)
- Pineal cell tumors including pineocytoma, pineoblastoma, and mixed tumor
- Other tumors including meningioma, astrocytoma, ganglioglioma, and dermoid cysts.

*Please see these individual tumor types for additional information.*

PINEAL TUMORS, PINEOCYTOMA, PINEOBLASTOMA, MIXED PINEAL TUMOR

These tumors originate from normal cells of the pineal gland—a gland located in the center of the brain involved in the secretion of specific hormones. The pineocytoma is a slow-growing, grade II tumor. Pineoblastoma is the more aggressive, grade IV, malignant counterpart. A grade III intermediate form has also been described. These tumors tend to occur in young adults between the age of 20 and 40 years. About 10 – 20 % of the tumors, particularly the pineoblastoma, have the potential to spread through the cerebrospinal fluid. This usually occurs late in the disease. The tumors, however, rarely spread elsewhere in the body.

Symptoms are most often due to obstruction of cerebrospinal fluid flow and involvement of the eye movement pathways. Headache, nausea and vomiting, and double vision are common.

CT and MRI scans are used to visualize the tumor and determine if it has spread. If a biopsy or surgery is not considered, cerebrospinal fluid analysis may be used to rule out other tumor types occurring in the same area.

Surgery may be possible in some individuals to determine the tumor type and to remove part of the tumor. If surgical removal is not possible, biopsy may be considered—depending on the risks involved—for purposes of obtaining a tissue sample for pathological examination and confirmation of the diagnosis. Some patients require placement of a shunt to relieve the cerebrospinal fluid obstruction.

The standard treatment for these tumors is radiation therapy. Conventional radiation therapy to the pineal region is standard, although a form of focused radiation may be considered. Radiation of the entire brain and spinal cord is generally recommended for pineoblastoma. Chemotherapy may also be considered, particularly if the tumor has spread or if the tumor regrows.

Pituitary Tumors

The pituitary gland is involved in the secretion of several essential hormones. Tumors arising from the pituitary gland itself are called adenomas. Adenomas are benign and slow growing tumors. These tumors represent approximately 10% of primary brain tumors. Pituitary adenomas occur at any age but incidence increases with age. Women are more affected than men, particularly during childbearing years.
Most pituitary adenomas grow in the front two-thirds of the pituitary gland, which is called the adenohypophysis. The tumors are classified as “secreting” or “non-secreting.” A “secreting tumor” produces excessive amounts of hormones. The majority of pituitary adenomas are secreting tumors, and are further classified by the hormone(s) being secreted.

Symptoms are caused by the growing tumor pushing on surrounding structures, by excessive hormone production, or by impaired hormone production. The hormones most commonly affected include growth hormone (which regulates body height and structure), prolactin (which controls lactation, or milk production), the sex hormones (which control the menstrual cycle and other sexual functions), thyroid gland hormones, adrenal gland hormones and vasopressin (which is involved in water and electrolyte balance). Pressure on surrounding structures most commonly causes headache, visual impairment, and behavioral changes.

MRI scan is used to determine the tumor size and position in relation to other brain structures. Several blood tests can be performed to determine which hormones are elevated or reduced.

Because hormones affect other parts of the body, treating a pituitary tumor is a team effort involving many specialists including a neurosurgeon, an endocrinologist and an ophthalmologist. Usually therapy includes surgery for tumor removal using an approach through the nose and sinuses. However, for a small prolactin-secreting pituitary adenoma, the drug, bromocriptine, may be used to reduce tumor size without surgery. Other tumor-shrinking drugs may be used after surgery depending on the type of hormone the tumor is secreting. Radiation therapy is used for a persistent or recurrent tumor that is not responding to drugs (if the tumor is secreting). For non-secreting tumors, radiation may be used following a partial removal or if the tumor was invasive. Replacement hormone therapy is often prescribed following surgery and/or radiation.

The pituitary carcinoma is the rare malignant form of the pituitary adenoma. It is diagnosed only when there is proven spread (metastases) inside or outside the nervous system. Symptoms are identical to those of the adenoma; this tumor may also secrete a variety of hormones. Treatment may include surgery, radiation therapy, hormone therapy, and chemotherapy.

**PNET—Primitive Neuroectodermal Tumor**

PNET is a name used for tumors which appear identical under the microscope to the medulloblastoma, but occur primarily in the cerebrum. PNET is used by some to designate tumors such as the pineoblastoma, polar spongioblastoma, medulloblastoma and medulloepithelioma. With the exception of the medulloblastoma, all of these are very rare tumors.

PNETs most frequently occur in very young children. The tumors contain undeveloped brain cells, are highly malignant, and tend to spread throughout the central nervous system.

PNETs commonly contain areas of dead tumor cells (necrosis) and cysts. Laboratory tests help differentiate this tumor from other types. CT scans can show cysts and areas of calcification which are common in PNETs. Surrounding edema is uncommon. MRI scans can provide an indication of tumor size.

Because they tend to be large tumors, symptoms of increased intracranial pressure and mass effect are usual. Seizures are common.

Surgery is the standard initial treatment for these tumors. Because of their large size and tendency to spread, as well as their extensive blood supply, total surgical removal is rarely achieved. In children older than three and in young adults, radiation therapy routinely follows surgery, with delivery to the entire brain and spine. Doses are similar to those used for medulloblastoma. Younger children are often treated with chemotherapy instead of radiation therapy, until they are older.

The chemotherapy used for medulloblastoma might also be effective against a PNET. Many clinical trials using chemotherapy and combinations of therapy are available for the medulloblastoma and PNET.
Pseudotumor Cerebri

This is an older term for a type of Primary or Idiopathic Intracranial Hypertension.

The cause of this condition is unknown, but it is not due to a brain tumor, and it is not a “true” brain tumor.

Pseudotumor cerebri literally means “false brain tumor” since there is increased pressure of the spinal fluid, which causes symptoms of increased intracranial pressure as may occur with a tumor. It is most common in young or middle-age individuals.

Headaches, vision changes, and other symptoms of increased intracranial pressure are usually present. Diagnosis is generally confirmed by a spinal tap which measures spinal fluid pressure. Scans are used to be sure an actual tumor does not exist.

Treatment consists of relieving the symptoms and saving vision. Pressure may be controlled by medications which reduce the production of cerebrospinal fluid. Vision will be closely monitored during treatment. If vision is threatened, surgery may be considered to decrease pressure on the optic nerves. Or, a shunt may be placed to move spinal fluid to another area of the body.

For more information on pseudotumor cerebri, contact:

Intracranial Hypertension Research Foundation
6517 Buena Vista Drive
Vancouver, WA 98661
Phone: 360-693-4473
Website: www.ihrfoundation.org

Schwannoma

See Acoustic Neuroma

Skull Base Tumors

Tumors located along the bones that form the bottom of the skull or along the bony ridge in back of the eyes are called skull base tumors. These tumors are most often chordomas, meningiomas, glomus jugulare, schwannomas or metastatic tumors.

Skull base tumors are diagnosed by MRI or CT scans. Treatment depends on the type and location of the tumor, and well as its surgical accessibility. Newer surgical tools and stereotactic technique help the neurosurgeon remove large portions of these tumors. A team approach might be used with extensive tumors: a neurosurgeon, an ear, nose and throat (otorhinolaryngology) specialist, a surgeon with training in cranio-facial surgery, and/or a plastic surgeon.

Outcome depends on the extent of tumor removal and the type of tumor. Recurrent tumors might be treated with a second surgery or focused radiation such as stereotactic radiosurgery. Radiation might also be used for partially removed or metastatic tumors.

Spinal Cord Tumors

The types of tumors found in the spine vary by location. Tumors commonly found on the outermost layer of the spinal cord lining, called the dura mater or the epidural area, include metastases from cancers which began elsewhere in the body as well as cancers which tend to be found near the spinal cord, such as sarcoma, neuroblastoma, and multiple myeloma. Tumors found within the dura mater but located outside the actual spinal cord substance (intradural/extradural) include meningioma,
schwannoma, neurofibroma, and other rare tumors. Finally, tumors arising within the spinal cord substance (intramedullary) include astrocytoma, ependymomas, and less commonly, metastases.

Some symptoms of spinal tumors are due to compression of the spinal cord and usually have a gradual onset. Pain and leg weakness are the most common. If the tumor infiltrates the spinal cord, pain is less common and weakness, sensory impairment, and bladder control difficulty are more often seen.

Treatment of spinal tumors depends on whether the tumor is primary or metastatic, its location and the type of tumor. Epidural tumors are usually treated with radiation with or without surgery. Surgery is the standard treatment for intradural, extramedullary tumors arising in the spinal canal. Intramedullary tumors may be treated with surgery and radiation may be indicated if residual tumor is left or if the tumor is high-grade.

**Teratoma**

A teratoma is characterized by the presence of the tissue types within the tumor—the tissue can be “mature” (developed cells) or immature (undeveloped). The mature teratoma is a rare, benign germ cell tumor which most frequently occurs in male infants and children. Teratomas represent 18-20% of all germ cell tumors. They often contain calcium, cysts, fat and other soft tissues.

Teratomas are most frequently found in the rear of the third ventricle near the pineal gland and above the pituitary gland. This germ cell tumor is the least likely to spread via the cerebrospinal fluid.

Teratomas are the most common brain tumor in newborns. The majority of teratomas occur in children and adolescents. The symptoms vary with the location (see sections on pineal region tumors and pituitary tumors). MRI scan and evaluation of the cerebrospinal fluid for tumor cells help determine if the tumor has spread.

Because obstruction of cerebrospinal fluid circulation occurs in most patients with pineal region tumors, a shunt is often needed to reduce the size of the ventricles before any other treatment can be performed. Surgery is the standard treatment for accessible tumors and can be curative. If surgery is not practical, a biopsy to establish an exact diagnosis may be possible. Radiation therapy may follow surgery or be used for inoperable or partially removed tumors particularly if immature. For children under the age of three, chemotherapy to delay radiation might be recommended.

**Tuberous Sclerosis Complex**

Tuberous sclerosis complex (TSC) is a hereditary disorder affecting the nervous system and the skin as well as other organs. It is an autosomal dominant genetic disorder meaning a child has a fifty-fifty chance of
inheriting TSC if his/her parent has this disorder. In addition, this disease may result from a sporadic mutation (the first case in a family) in 50–60% of individuals.

TSC may be diagnosed at any time in an individual's life depending on the onset of symptoms and the severity of the disease.

TSC causes tumors to form in several parts of the body. The brain tumor associated with this disease is called a subependymal giant cell astrocytoma (SEGA). It often occurs near the foramen of Monro (the narrow channel, also called the interventricular foramen, between the lateral ventricles and the third ventricle through which cerebrospinal fluid flows). This tumor occurs in 5-20% of individuals with TSC.

Surgery is generally recommended when tumors show signs of continued growth. Current clinical trials with a specific group of drugs called mTOR inhibitors may reduce the future role of surgery for these tumors if these treatments prove to be successful. Radiation and chemotherapy are contraindicated for this type of tumor.

Diagnosis, screening and follow-up care for individuals with TSC are available through TSC clinics.

For more information on TSC, contact:

Tuberous Sclerosis Alliance
801 Roeder Road, Suite 750
Silver Spring, Maryland  20910

Phone: 800-225-6872   or 301-562-9890
Website: [www.tsalliance.org](http://www.tsalliance.org)

**Vestibular Schwannoma**

See Acoustic Neuroma
Seizures are common symptoms of a brain tumor. 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. During abnormal electrical activity, something interferes with those signals and they become more intense. The result is a seizure.

Seizures are usually brief. However, their effects may linger for several hours. Recurrent seizures are referred to as epilepsy.

People with seizure activity may experience:
- Unusual movements
- Change in the level or loss of consciousness
- Sensory distortions

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

**Seizure Warning Signs**
Most seizures occur randomly, at any time, and without any particular cause. However, you might have some advance notice. Learning these signals—called auras—can help you prepare for a seizure, and help you care for yourself or your loved one.

An aura may happen a few seconds or minutes before the actual seizure. Headache, mood changes, and/or muscle jerking might signal a coming seizure. Use that time to safeguard yourself. For example, if you are chewing, remove the food from your mouth. If you are walking, sit or lie down. If are with someone experiencing an aura, assist them in finding a safe place.

Some individuals use service dogs that are able to detect an approaching seizure and can then guide their owner to safety. For more information on service dogs, visit the Delta Society’s Web site at: www.deltasociety.org

The American Brain Tumor Association also offers Emergency Alert Wallet cards, which can be obtained by calling us at 800-886-2282, or sending an e-mail to: info@abta.org.

**Seizure Triggers**
If you have recurrent seizures, you might notice that some events “trigger” them. Bright lights, flashing lights, loud noises, specific odors, lack of sleep, missed meals, menses, increased stress or emotional difficulties, alcohol, new medications, or changed dosages of existing medications can all be triggers. Keeping track of what you were doing immediately prior to each seizure can help you identify your personal triggers.

**Types of Seizures**
There are different types of seizures. The type you experience depends on which area of the brain has the abnormal electrical signals.

There are two primary types of seizures.
- Partial seizures
- Generalized seizures

**PARTIAL (ALSO CALLED “FOCAL”) SEIZURES**
There are two types of partial seizures.
- Simple partial seizures
- Complex 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...
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. More information about these medications is offered further into this chapter.

**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, lower 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 the diet could be followed by adults, AEDs tend to be prescribed first due to the diet’s very strict food restrictions.

### 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 caring presence and observation.

First, make sure the person is breathing. Loosen clothes around the neck. 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, take a moment to clear the area of sharp objects or anything else that could be dangerous. 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.

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

About Antiepileptic Drugs (AEDs)

The goal of antiepileptic drugs (AEDs), also called seizure medications, is to prevent seizures with the lowest effective doses and the least side-effects.

Maintain a Steady Level
- Seizure medications work best when there is a steady level of the drug in your body. They need to reach and remain at the ideal level to be effective.
- Remember to take your medication regularly as prescribed. If you miss a dose, do not double up. Resume your regular schedule and notify your doctor. If you stop taking your medicine abruptly, seizure activity will increase. If you miss more than one dose, notice an increase in your seizures, or develop a rash, call your doctor for assistance.
- Do not change the dosage or stop taking your medicine without the approval of your doctor. If one medication does not control your seizures, another drug or a combination of drugs may be prescribed.

Check Levels if Indicated
- Some seizure medications require frequent blood tests to check the drug levels. Ask your doctor if the medication you are using needs to be monitored in this way and where and when to have those blood tests done. Your medications might be adjusted based on the results.

Minimize Possible Drug-Interactions
- There are many medications, both prescription and over-the-counter, which can influence the effectiveness of seizure medications. Be sure your doctor is aware of all the medications you take. Do not forget to mention vitamin and nutritional supplements, or herbal medications you may be using.

Ask About The Guidelines
- Talk with your doctor and nurse about your risk for seizures, and their guidelines for keeping you on the AEDs.
- Your need for AEDs is based on your seizure history, your MRI scans, EEG (electroencephalogram) results, and your treatments. The decision to taper off seizure medications should be carefully planned with your doctor. Never stop your AEDs without consulting your healthcare team.
**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. Information about side-effects of specific AEDs can be found at the ABTA web site. Please visit us at www.abta.org.

**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**

This side-effect occurs from some seizure medications. Your gums may be inflamed, red, swollen, tender, and 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 mouth care “sponge” available at most drug stores. A soft toothbrush is another option. 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**

Be sure to 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. 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 arouse 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 for me?
- 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?

Other Questions I would like to ask:
__________________________________
__________________________________
__________________________________
__________________________________
__________________________________
__________________________________
__________________________________

Living with Seizures

Seizures are generally unpredictable. It is important to remember that seizures can be managed, and you can influence how you feel about your seizure disorder. Here are a few suggestions for putting control back in your hands:

Stress

Uncertainty can take its toll physically, emotionally, and socially. Aim to be gentle with yourself. Exercise, meditation, yoga, guided imagery, deep breathing, and coping skills training are all ways in which you can support yourself, and potentially reduce stress.

Relationships

Yes, a seizure disorder can be stressful to you—and it 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 a seizure disorder 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

While our society is heavily dependant on transportation systems to move us around, remember that laws prohibiting people with seizures from driving are designed to protect both you and other people from injury. Talk with a Social Worker 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.
Related ABTA Resources & Publications

- Reaching Out for Support for support groups, pen pal programs, counseling, neuropsychologists, and telephone support
- Therapeutic Recreation for services to improve quality of life
- Neuropsychology for assistance with defining and understanding cognitive skills, psychological functioning, and behavior
- Transportation Resources for resources and information about transportation support
- Working with a Brain Tumor for employment rights, job retraining, legal rights, and more
- Financial Assistance Resources for resources and information about financial support
- Care for the Caregiver Corner for support groups, pen pal programs, orientation to caregiving handbook, online support groups and websites, resources for stress management and self-care, and other resources

For More Information About Seizures, Living with Seizures, and Medications, contact:

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<th>American Epilepsy Outreach Foundation</th>
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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 even 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.

We hope some of these ideas are helpful for you.
Understanding Your Disease

For many people, learning and understanding is the foundation for coping. By becoming more knowledgeable and aware, you will be able to make informed medical decisions to the best of your ability. As a result, this will help you gain a greater sense of control over your body and your medical care.

Although it will be difficult to process everything, begin by listening carefully to your doctors and nurses when they explain your illness and treatment options. They are your best source for information about your brain tumor. Don’t be afraid to ask questions, repeat words and pronunciations or have your diagnosis spelled out for you. Most healthcare professionals want you to have accurate information so you can actively manage your treatment.

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, he/she 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 okay 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. 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.

Telling Family and Friends

Telling your family and friends that you have a brain tumor can be difficult. If you are uncomfortable talking with your family, consider having a care conference or meeting with your doctor, health care team and the primary members of your family. Talk with your nurse or doctor if you think a care conference would be helpful for you and your family.

Also, 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

**TALKING WITH CHILDREN**

These sample explanations can be adapted for conversations with your children. Change the phrases to match your situation.

“The doctor wants to do some tests to find out why I am getting sick to my stomach and having headaches…”

“An MRI scan takes picture of your 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.”

“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. We also know that you don’t ‘catch’ brain tumors from other people.”

“Would you like to talk about this? Is there anything you would like to ask?”

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

For additional explanations children can understand, please visit our children’s web site—ABTA Kids—at www.abta.org/kids/home.htm or request a free copy of our DVD, Alex’s Journey.
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 ABTA, community centers, social service agencies, government health agencies and schools.

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. Young people often have remarkable insight and can be a source of great comfort.

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. A list of books is available through ABTA.

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.

If friends offer to help, accept their offers. You will benefit from the assistance, and your friends will feel needed. Groceries, laundry, 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. Don’t be shy! Or create an online calendar with tasks that need to be done. Friends and family members can log on to one of the many web based caring sites and sign up to help with a need.

Although most people will be supportive, some may be unable to deal with or even acknowledge your illness. 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.

Your Feelings

When you first heard about your diagnosis, you were probably shocked. Chances are you may not have understood or remember what you were told at that time. This is a normal reaction. Most people experience some or all of the following emotions after the diagnosis of a brain tumor:

DENIAL OR SHOCK
Denial—disbelief or lack of concern over the diagnosis—is normal for some. It may take time to accept the news. Some may initially pretend it hasn’t happened. Others may be in a state of shock. “How could I have a brain tumor?” or “Why me?” are common questions. Some people may refuse to discuss or even acknowledge their diagnosis.

GUILT
When something overwhelming happens, people try to blame someone. When you blame yourself, you feel guilt. People ask: “Is this a punishment?” or “Did I do something to deserve this?” The cause of most brain tumors is unknown. Nothing you did, said or thought made this happen.

ANGER
Anger at your husband, wife, children, neighbor, boss, doctor or anyone and everyone—is not unusual. You may say hurtful, bitter things that you don’t mean and later regret. Small children may kick or bite to show their anger. Hidden or repressed anger sometimes causes irritability, sleeplessness, fatigue, over-eating, or over-drinking.

DEPRESSION
Depression or grief at the loss of your previous lifestyle may occur. 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 first must be diagnosed.

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.

There is no magic pattern for dealing with your emotions. One day you may feel better, while the next day you may feel uneasy again. Not everyone shows their emotions, nor does everyone have the same kinds of 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.

Living Your Life

Part of our identity is how we present ourselves to others. An undesired change in the way we look can understandably be upsetting.

Hair lost during surgery, radiation, or chemotherapy often grows back, but may take months. Wigs are available for both men and women. If you find a wig to be uncomfortable, consider a scarf or a loose hat. A listing of wigs and head coverings is available through ABTA.

Look through your closet for the clothes you look best in. Or, treat yourself to a new blouse or tie. Oftentimes, when you look good, you feel better. Many hospitals offer make-up and hair sessions for those who have gone through cancer treatments. These sessions offer tips about enhancing your appearance which can be helpful for improving your self-confidence.

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

You may feel tired due to medications, treatments, and traveling to and from your treatments. Be realistic—keeping up with your usual responsibilities may be too difficult. 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.

Make time to be good to yourself. Take up a hobby or learn a new craft. Go to the library and check out those books you always wanted to read. Keep a journal, take a walk, pray or laugh. Look for ways to enjoy yourself.

Coping With 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 soft music, journaling, reading a book or taking a mid-afternoon nap—all are 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 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.

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.

**Finding a Support Group**

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. Due to this, there is often a need to connect with others in similar situations.

Patients and families often find help through brain tumor support groups. A support group is a 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. Within the safety of a support group, many people are able to share their fears and concerns about day-to-day problems and the future.

There are different types of support groups for adults, parents of children with brain tumors, children and siblings. Most of these groups also welcome concerned friends. 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 is a lot like the process of trial and error.

**SOCIAL WORK SERVICES**

The American Brain Tumor Association social workers can help you or your family explore a wide variety of support options. There are many sources of online support, community-based programs, and opportunities to contact others living with this diagnosis. Our Social Workers can be reached at 800-886-2282 or socialwork@abta.org
Whether you are just beginning treatment, are a long term survivor, or 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 healthcare team. Your doctors and nurses can respond with personalized answers which cannot—and often should not—be provided by outside sources. By asking questions you’re participating in your healthcare. 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.
Talking to Your Healthcare Team

Make a list of your questions and bring them with you when you visit your doctor. Be sure the questions that concern you the most are at the top of the list. If you think of other questions after you return home, begin a list for the next visit.

If you want to bring family or friends with you when you visit your doctor, make an appointment for a conference. Let the receptionist know the purpose of your visit — that way, a block of time can be set aside for you.

Your First Visit

Many people don’t remember much when their doctor first tells them they have a serious disease. Try to come away with some basic information:

- Where is the tumor located?
- Based on the scans, do you have an idea of the type of tumor?
- What is the next step? Do I need more tests? Do I need to see any specialists?
- Until we know more, should I continue my daily routine? Can I work? Should I drive?
- Do I need to take any medication? If so, what is it for? What are the side-effects?

About Insurance

After your first visit, you’ll need to verify your healthcare insurance coverage. The answers to most of your insurance questions can be found in the insurance policy itself or the policy manual. If you 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 800-633-4227. For CHIP coverage (Comprehensive Health Insurance Programs) through your state, call your state Department of Insurance.

If you are uninsured, please 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 healthcare coverage.

Questions for Your Insurance Provider

Be sure you know the answers to these questions:

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

QUICKEN 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?

Following Treatment…
A Next Step
Eventually, the frequent appointments for therapy stop, and the dates for follow-up care become further apart. The pace slows, and another period of adjustment begins. Now is the time to begin re-defining “normal” in your life. It’s a time to slow down and be good to yourself.

QUESTIONS FOLLOWING TREATMENT
When you finish your last treatment, be sure you know:
- When is my next doctor visit? Which doctor(s) do I see, and how often?
- When is my next scan? Do I need a doctor’s order? How will it be scheduled?
- Do I need any medications? If so, are there any potential side-effects?
- Can I work? If so, do I have any restrictions on my activities? Will I need any work site accommodations?
- Can I drive?
- Can I exercise? If so, do I have any limitations?
- What type of diet should I follow? Is there a registered dietician I can consult to provide me with healthful eating guidelines?

Living with a Brain Tumor
As time goes on, you and your family may have questions about concerns common to all people living with a brain tumor — patients and family members alike. Those concerns may involve:
- financial assistance
- employment interests
- vocational re-training
- obtaining new health insurance
- sexuality
- forming new relationships
- starting or adding to your family
- parenting
- counseling or support groups
- rehabilitative services
- cosmetic and image interests

As more brain tumor patients become survivors, there are increasing resources for people living with a brain tumor. Tap into these services and learn how they can help enrich your life. A social work program can help you explore these, and other, opportunities. Please feel free to call us at 800-886-2282.
Please ask a member of your healthcare team to complete the form on page 75.

Use it to learn the exact spelling of your tumor type, and its location, your medications, and resources for additional information.
The name of my tumor is:

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

My nurse’s name is: __________________________
Phone: _________________________________

For information about brain tumors:

American Brain Tumor Association (ABTA)
www.abta.org 800-886-2282

Cancer Information Service (CIS) of the National Cancer Institute (NCI)
www.cancer.gov 800-422-6237

National Institute of Neurological Disorders & Stroke (NINDS)
www.ninds.nih.gov 800-352-9424

Where is my tumor?

Lobes of the Brain

When is my next appointment? With whom?

I take these medications:

The American Brain Tumor Association exists to eliminate brain tumors through research and to meet the needs of brain tumor patients and their families.
Central Nervous System

American Brain Tumor Association • 800-886-2282
BUILDING KNOWLEDGE
Dictionary for Brain Tumor Patients
Living with a Brain Tumor
A Primer of Brain Tumors

FOCUSING ON TUMORS SERIES
Ependymoma
Glioblastoma Multiforme and Anaplastic Astrocytoma
Medulloblastoma
Meningioma
Metastatic Brain Tumors
Oligodendroglioma and Oligoastrocytoma
Pituitary Tumors

FOCUSING ON TREATMENT SERIES
Chemotherapy
Conventional Radiation Therapy
Stereotactic Radiosurgery
Steroids
Surgery
Physician Resource List: Physicians Offering Clinical Trials for Brain Tumors

FOR & ABOUT CHILDREN
Adolescent and Young Adult Resources
Alex’s Journey: The Story of a Child with a Brain Tumor (DVD)
Educating Children and Teenagers
Resources for Talking with Children When a Parent is Ill
When Your Child Returns to School

SUPPORT RESOURCES
Bibliography
Care Options
Caregiver Resources
Emergency Alert Wallet Cards
Employment Rights and Job Retraining Resources
End-of-Life Care
Financial Assistance Resources
Health Insurance Resources
Housing During Treatment Resources
Neuropsychology Resources
Scholarship & Educational Financial Aid Resources
Searching Medical Journals
Social Security Disability Resources
Spanish-Language Resources
Therapeutic Recreation Resources
Transportation Assistance Resources
Wig and Head Covering Resources
Wish Fulfillment Resources

NEWS
Headlines
ABTA Brain Tumor E-News

FOCUSING ON SUPPORT SERIES
Listing of Brain Tumor Support Groups
Listing of Bereavement (Grief) Support Groups
Organizing and Facilitating Support Groups
Pen Pal Programs
• Connections (for patients and family members)
• Bridges (for those who have lost someone to a brain tumor)
Reaching Out for Support
Resources for Online Support
TLC (Tips for Living and Coping) e-bulletin

Single copies of these publications are available free of charge.

American Brain Tumor Association
2720 River Road, Suite 146 Des Plaines, Illinois 60018
800.886.2282 TEL info@abta.org EMAIL
847.827.9918 FAX www.abta.org WEB