



FOCUSING ON TUMORS



American Brain Tumor Association

## A Word About ABTA

Founded in 1973, the American Brain Tumor Association was the first national, nonprofit organization dedicated solely to brain tumors. Since then, ABTA has been a consistent leader in brain tumor research, and patient/family information and support.

We gratefully acknowledge Regina Jakacki, MD, Director, Pediatric Neuro-Oncology Program, Children's Hospital of Pittsburgh, Pittsburgh, Pennsylvania, for this edition of "Ependymoma." We also thank Beverly LaVally, RN, of the Dana-Farber Cancer Institute, Boston, for coordinating the original edition of this publication.

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## Introduction

Any tumor that arises from the glial, or supportive, tissue of the brain is called a "glioma." One type of glioma is the ependymoma. Ependymomas arise from ependymal cells that line the ventricles of the brain and the center of the spinal cord. Occasionally, ependymal cells are found within the brain itself. Ependymomas are soft, grayish or red tumors which may contain cysts or mineral calcifications.

### *Ependymal cells*

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



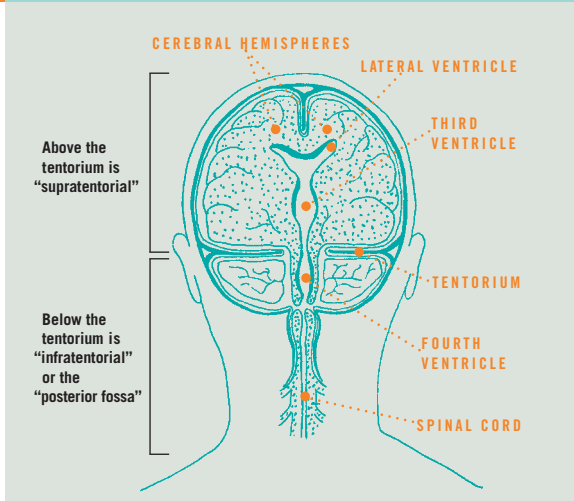
The words "supratentorial" and "infratentorial" describe the location of the ependymoma within the brain. The tentorium is a thick membrane that separates the top two-thirds of the brain from the bottom third. Supratentorial ependymomas occur above the tentorium.

This area includes the cerebral hemispheres, as well as the lateral and the third ventricles. Infratentorial ependymomas occur below the tentorium. This area, also called the posterior fossa, includes the fourth ventricle, the brainstem and the cerebellum.

There are four major types of these tumors: myxopapillary ependymomas, subependymomas, ependymomas, and anaplastic ependymomas.

Many pathologists also assign a number “grade” to ependymomas. The grade is based on how much the cells look like normal ependymal cells, although various grading systems exist. While the cells of a grade I tumor look somewhat unusual, grade III tumor cells look definitely abnormal.

### The Tentorium



Most grade I tumors do not recur after complete surgical removal. Grades II and III tumors need additional treatment and have a higher likelihood of recurring.

Myxopapillary ependymomas tend to occur in the lower part of the spinal column. Subependymomas usually occur near a ventricle. Both 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.

Anaplastic ependymomas are high-grade tumors (grade III) and tend to be faster growing than low-grade tumors. They most commonly occur in the posterior fossa.

### Incidence

Ependymomas are relatively rare tumors, accounting for 2-3% of all primary brain tumors. However, they are the fourth most common brain tumor in children. About 30% of pediatric ependymomas are diagnosed in children younger than 3 years of age.

The location of ependymomas in adults tends to be different than the location of ependymomas in children. In adults, 60% of these tumors are found in the spinal cord. In children, 90% of ependymomas are found in the brain, with the majority located in the posterior fossa.

### Cause

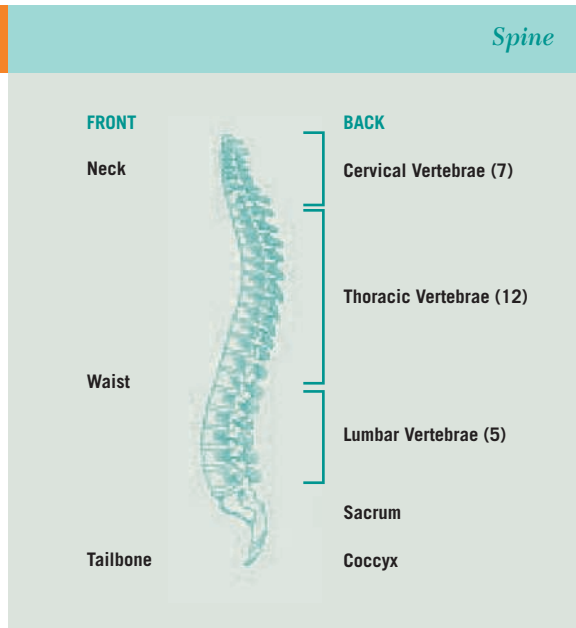
The cause of ependymomas, like that of other brain tumors, is unknown. It is uncertain whether viruses (e.g. SV40) play a role in the development of ependymomas. Much more research is necessary to determine whether this is indeed a factor in humans, since these particular viruses do not normally affect humans.

### Symptoms

Symptoms of an ependymoma are related to the location and size of the tumor. In neonates and infants, enlargement of the head may be one of the first symptoms. Irritability, sleepiness and vomiting may develop as the tumor grows. In older

children and adults, nausea, vomiting and headache are the most common symptoms.

These are usually signs of increased pressure, which may develop if the tumor blocks the drainage of cerebrospinal fluid (the liquid that bathes the brain).



Headaches are a common symptom, and are generally worse in the morning. A tumor near the brainstem may cause one or both eyes to cross, balance problems, or trouble walking.

Neck pain may result from a tumor growing near the brainstem or the upper part of the spinal cord. Seizures, headaches and weakness of one side or one part of the body may occur with a tumor in the cerebral hemispheres. Spinal cord tumors often cause leg or back pain which can be severe enough to awaken the person from sleep. Tingling sensations, numbness, or weakness in the arms or legs may also occur. Difficulty with bladder or bowel

control may be caused by an ependymoma in the lower part of the spine.

If you would like to know more about the symptoms of brain tumors, please visit our Web site at [www.abta.org](http://www.abta.org).

## Diagnosis

Magnetic Resonance Imaging (MRI) scans and/or Computerized Axial Tomography (CT) scans are required for patients suspected of having a brain tumor. MRI scanning, which uses radio waves and a magnetic field, provides details about the location of the tumor and which parts of the brain or spinal cord are involved prior to surgery. CT scanners are sophisticated x-ray devices connected to computers. CT scans are useful for getting a quick view of the brain to see if there is a tumor and



Photo courtesy of Dr. Jakacki

whether it has caused hydrocephalus (the buildup of cerebrospinal fluid in the brain). However, only microscopic examination of a sample of tissue obtained during surgery or biopsy confirms the exact diagnosis.

About 10-15% of ependymomas spread, or metastasize, through the cerebrospinal fluid. The tumor cells then may grow independently, in or along the spinal cord, or rarely, in other places in the brain. Infratentorial tumors are more likely to spread to the spine than supratentorial tumors. Ependymomas almost never spread outside of the brain and spinal cord. A spinal MRI scan and lumbar puncture (spinal tap) will be performed to determine whether the tumor has spread to the spine and/or spinal fluid. The fluid obtained during the spinal tap will be tested for the presence of tumor cells. Your doctor will decide the appropriate time to perform these tests. The results are used to guide treatment.

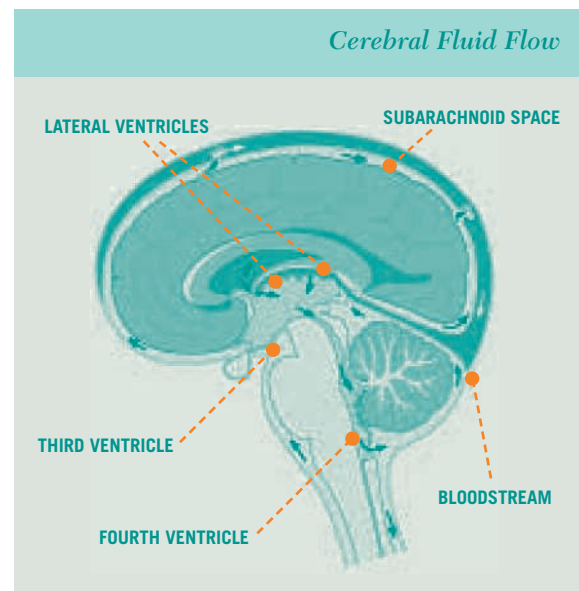
## Treatment

The treatment of an ependymoma varies depending on its location, grade, and whether the tumor has spread to the spine.

## Surgery

The most important step in the treatment of an ependymoma is surgery to remove as much tumor as possible. Studies clearly show that patients whose tumor can be “grossly” removed (removing all tumor that can be seen) have the best chance of long-term survival. The amount of tumor that can be removed, however, depends on the location of the tumor. High-powered microscopes in the operating room help the surgeon see

tumor located in and around the ventricles or the brainstem. Removal of all visible tumor is not always possible, especially if the tumor is attached to the brainstem or involves other important areas of the brain. In children, ependymomas often fill the fourth ventricle and extend through its floor into the brainstem, making safe removal of those portions of tumor difficult.



Hydrocephalus is often present at the time of diagnosis if the tumor has blocked the out-flow of cerebrospinal fluid. Tumor removal may open up these fluid pathways, although some patients will still require the placement of a ventriculoperitoneal (VP) shunt to by-pass the blockage. Many surgeons will place a temporary external ventricular drain during surgery. This is used to drain off excess fluid for a few days after the surgery. If it cannot be removed after a few days, a permanent shunt may be required.

An MRI of the brain should be done within a day or two of surgery to determine how much visible tumor remains, if any.

### **Radiation**

Radiation therapy is usually recommended for older children (age dependent on the location of the tumor) and adults following surgery, even if all visible tumor was removed. Patients treated with radiation therapy following surgery generally have a better chance of long-term survival than patients treated with surgery alone.

The age of the patient, the location of the tumor, how easily it was removed, and whether it has spread are considered in treatment planning.

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.

There are different methods of administering radiation. External beam radiation is given 5 days a week for 6 weeks. Conformal beam radiation therapy is a type of external beam radiation that contours the radiation beams to the shape of the tumor. Radiosurgery is a way of giving a single high-dose of precisely focused radiation to the tumor. This is often used for ependymomas that grow back after conventional radiation. Your doctor will decide which form of radiation is best for you.

Because of the long-term side effects of radiating young children, chemotherapy may be used to delay radiation therapy in the very young child. If the tumor grows despite treatment with chemotherapy, radiation therapy may be considered for these children.

### **Chemotherapy**

Chemotherapy uses special drugs to kill tumor cells. Scientists continue to study the role of chemotherapy and when it is best used. It is not clear whether using chemotherapy in the treatment of patients with newly diagnosed ependymomas makes a difference. Some tumors respond to treatment for awhile, while others continue to grow. Chemotherapy may also be used to delay radiation for infants and young children, or to treat tumors that have grown back after radiation therapy. It is not clear which chemotherapy drugs are the most effective against ependymomas, although cisplatin and carboplatin can cause shrinkage in about half of ependymomas, although not usually for a long time. Either standard chemotherapy, or experimental chemotherapy as part of a clinical trial, are often used for patients whose tumors regrow after radiation.

### **To Learn More**

With improved neurosurgical and radiation therapy techniques, patients with ependymomas have a significantly increased chance of survival, particularly if they are cared for by an experienced multi-disciplinary team of medical professionals at an established pediatric or adult cancer center.

Additional treatment information about this tumor is available from the National Cancer Institute's Cancer Information Service at 800-422-6237. Choose the option of speaking to an information specialist. CIS can provide information about ependymomas in adults or children, and a list of clinical trials (investigative new treatments) open to those with an ependymoma. Be sure to share the information you find with your healthcare team. They can advise you about the benefits and risks of the treatments based on your individual tumor. The American Brain Tumor Association maintains a listing of physicians involved in clinical trials, and extensive resources for support services. We can be reached at 800-886-2282.

### Treatment Side Effects

Some treatment, while increasing survival, may have significant side effects on children. Prior to treatment your child may undergo neuropsychological testing. Those results can be used as a baseline for future evaluations.

Surgery, particularly for infratentorial ependymomas, may result in the development or temporary worsening of balance, speech, or swallowing difficulties. Most of these changes will improve over several weeks or months.

The negative effects of radiation therapy on the brain are of great concern to those who treat children with brain tumors. A significant number of children, particularly those who are very young at the time of treatment, experience some degree of decreased intellect and learning problems

following radiation to large areas of the brain. The severity of the learning problems is correlated with the location and amount of brain radiated and the age of the child. Radiation to just the back part of the brain does not cause as many problems as to the upper part of the brain. When the upper part of the brain is radiated, teachers and parents often report significant problems with reading, writing, and short term memory. The role of special education programs to help these children develop their strengths and identify their weaknesses is of prime importance.

Older children and adults tend to have fewer problems. Poor growth may be a consequence of radiation therapy damage to the hypothalamus or pituitary gland which produce several important hormones. Replacement of these hormones is often necessary, under the supervision of a pediatric endocrinologist. Radiation to the spine at a young age may also cause short stature. Weakening of the muscles next to the spine is also possible.

The long-term effects of chemotherapy in children are still being studied. Platinum-based drugs often cause hearing loss, as can radiation therapy when delivered near the ears. Both radiation and chemotherapy can increase the chance of developing a second cancer or brain tumor. Some chemotherapy can cause infertility.

*The risk of incurring long term side effects due to treatment must be weighed against the outcome if these therapies are not administered. Your doctor can help you look at these concerns and understand the risks based on your child's particular treatment plan, as well as help you balance the benefits of therapy against the potential risks.*

## Follow-up

MRI scanning of the brain and/or spinal cord is usually done every 3-4 months for the first two years following diagnosis. These scans are used to determine the effectiveness of treatment and to watch for possible recurrence. Scans are done less frequently thereafter, unless symptoms develop which might indicate tumor growth. Your doctor will determine the appropriate schedule.

## Recurrence

The extent of tumor removal continues to be the strongest factor influencing recurrence and survival. Age at diagnosis, amount of tumor remaining after surgery, whether the tumor has spread, and the therapy given can all influence outcome. Researchers are also studying the biologic features of these tumors to determine which may be useful in predicting prognosis.

For patients whose tumors recur after initial treatment, re-operation when possible, followed by additional therapy, can be helpful. Newer investigational treatments include various forms of chemotherapy, and re-irradiation (possibly with radiosurgery) to the site of tumor recurrence.

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

## A Next Step

Our web site — [www.abta.org](http://www.abta.org) — offers extensive brain tumor information, treatment and research updates, patient/survivor/family stories, and a separate section for children called ABTA Kids.

A pen-pal program, Connections, links people interested in this disease with others. Parent/teacher education packets help ease your child's return to their classroom.

*Alex's Journey* is a fictional story of a child with a brain tumor.

Available in VHS or DVD format, this short movie is appropriate for a child with a brain tumor, siblings, or for use in a classroom. Our memory retraining, rehabilitative medicine, and employment resources may be of help to adult survivors and their families. Please call us at 800-886-2282 or visit our web site to access any of these programs.

The thread that runs through each of our services and programs is hope. Become involved — join us in some way to move towards a cure, and ultimately, prevention of brain tumors.

We hope that the information in this booklet helps you communicate better with the people caring for you or your child. Our purpose is not to provide answers; rather, we encourage you to ask questions.

## ABTA Publications and Services

### BUILDING KNOWLEDGE

Dictionary for Brain Tumor Patients  
Living with a Brain Tumor  
A Primer of Brain Tumors

### FOCUSING ON TUMORS

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

### FOCUSING ON TREATMENT

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

*Single copies of our publications are  
available free of charge.*

### 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

ABTA Headlines  
ABTA Brain Tumor E-News

### FOCUSING ON SUPPORT

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



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