

AMERICAN BRAIN TUMOR ASSOCIATION

Ependymoma



American
Brain Tumor
Association®

Providing and pursuing answers®

ABOUT THE AMERICAN BRAIN TUMOR ASSOCIATION

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To learn more, visit abta.org

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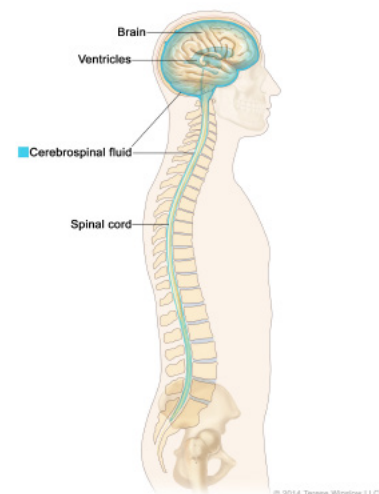
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Ependymoma

INTRODUCTION

This brochure is about **ependymoma** (pronounced ep-en-dy-MO-muh), a primary central nervous system (CNS) tumor, meaning that it begins in the brain or spinal cord and rarely spreads to other organs.¹ Ependymomas arise from ependymal cells, which form the lining of the **cerebrospinal fluid (CSF)** spaces in the CNS. Ependymomas can form anywhere in the CNS, but they often occur in the cerebellum and spinal cord.^{2,3} Sometimes, ependymomas can spread through the CSF to other areas of the brain, spine, or both.



Cerebrospinal fluid (CSF; shown in blue) is made by tissue that lines the ventricles (hollow spaces) in the brain. It flows in and around the brain and spinal cord to help cushion them from injury and provide nutrients. (cancer.gov)

TUMOR SUBTYPE

The World Health Organization (WHO) classifies primary CNS tumors into different types. **Ependymomas are classified into five subtypes.** They are:

- Subependymoma
- Myxopapillary ependymoma
- Classic ependymoma
 - Clear cell ependymoma
 - Papillary ependymoma
 - Tanycytic ependymoma
- Ependymoma, RELA fusion-positive
- Anaplastic (meaning cancerous) ependymoma

In recent years, researchers have learned a lot about ependymoma subtypes, in large part, due to advances in genetic testing (also referred to as molecular testing). They have learned that these subtypes are distinct and usually require different treatments.⁵ Researchers continue to identify new subtypes.^{6,7} In the future, this knowledge can lead to new therapies based on tumor subtype.^{8,9}

TUMOR LOCATION

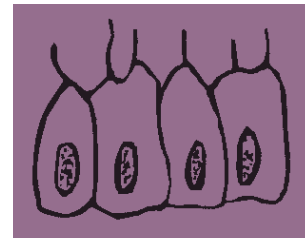
The location of a tumor is linked to the symptoms a person may have because the lobes of the brain and spinal cord control different functions, such as thought and reasoning, language, vision, hearing, movement, sensation, and bladder and bowel control.

In children, most ependymomas occur near or in the cerebellum^{1,2} - the part of the brain that controls balance and movement. It is estimated that as many as 90% of the ependymomas in children arise in the brain.^{6,10} **In contrast, ependymomas in the spine are more common in adults.^{1,6}** About 75% of ependymomas in adults are spinal tumors.^{6,10} It is important to understand that not all ependymomas arising from the cerebellum or the spinal cord behave

the same way or need the same treatment as tumors of different grades and subtypes can occur in the same location.

TUMOR GRADE

The WHO uses a grading system with a scale of I to IV for brain tumors; grade I is the mildest and grade IV is the most serious. The grade describes how much the tumor cells look like healthy cells or abnormal cells and how quickly the tumor is likely to grow and spread.^{2,3} Grade I and grade II tumor cells look more like normal cells and tend to grow more slowly than grade III and grade IV tumors.



Ependymal cells

Ependymomas are classified into grades I to III.^{1,4}

Tumor grading is important because it helps the doctor select the best treatment options. The WHO classification for ependymoma is:⁴

Grade I

- Subependymoma
- Myxopapillary ependymoma

Grade II

- Classic ependymoma
 - Papillary ependymoma
 - Clear cell ependymoma
 - Tanycytic ependymoma
- Ependymoma, RELA fusion-positive

Grade III

- Anaplastic ependymoma
- Ependymoma, RELA fusion-positive

Researchers are looking to find better ways to define the features that set a grade II ependymoma apart from a grade III ependymoma.² For example, the RELA fusion-positive subtype can be either a grade II or grade III tumor. Currently, it is unclear how the differences between grade II and grade III ependymomas affect a patient's prognosis (chance of recovery).⁶

Finding out the ependymoma subtype, as well as its location and grade, may help you make more informed treatment decisions.

INCIDENCE

Ependymomas are relatively rare, accounting for less than 2% of all brain and other CNS tumors.^{11,12}

Ependymomas can occur in both children and adults,^{1,2} but they occur more often in children.²

Ependymomas account for about 9% of all childhood brain and other CNS tumors.^{13,14} While the mean age of diagnosis is 4 years old, between 25% and 40% of patients are younger than 2 years of age.^{6,10} Incidence, which refers to how often a disease occurs, drops in pre-teens and teenagers.¹³ Ependymomas occur slightly more in males than females.¹⁵ They occur more commonly in white and non-hispanic people.¹

An estimated 1,430 people are diagnosed with ependymoma each year in the U.S.¹⁶ An estimated 13,294 people are living with ependymoma.¹

CAUSES

Like other types of brain tumors, **the exact cause of most ependymomas is unknown.**^{1,17} Researchers have identified abnormalities in the genes of different chromosomes that may play a role in how CNS tumors develop. But what causes normal CNS cells to change into abnormal tumor cells is still unclear.¹⁸

There are some risk factors that can increase a person's chance of developing a brain or CNS tumor. Risk factors don't directly cause the tumor to develop, but often influence the development of it. This explains why some people with many risk factors never get a tumor, while others who don't have any risk factors do develop a tumor.

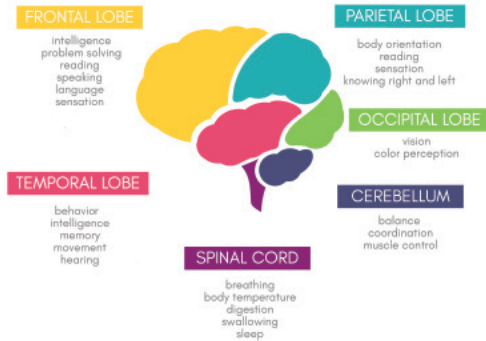
One known risk factor for developing ependymoma is an inherited cancer syndrome called neurofibromatosis type 2.^{6,17} However, people who inherit the genetic changes that cause neurofibromatosis type 2 do not inherit ependymoma, meaning that the risk of developing the tumor is not passed down to family members. Only a small number of patients have this cancer syndrome that increases their chance of developing ependymoma and not all patients with this syndrome develop ependymoma.

SYMPTOMS

As with other primary CNS tumors, **symptoms associated with ependymoma often depend on where the tumor is located** in the brain or spine.¹ Each person will experience unique or different symptoms due to the difference in location of the tumor.

Oftentimes, an ependymoma blocks the normal flow of CSF, which may increase pressure on the brain⁶ (also called hydrocephalus) or disrupt connections between normal brain cells. **Common symptoms** may include headaches or hydrocephalus; nausea and vomiting; dizziness; vision changes; seizures; personality changes;

weakness or numbness and tingling on one side of the body; and cognitive problems related to thinking, learning, concentrating, problem-solving, and decision-making.^{1,2}



A tumor in the spine may cause back and/or neck pain; numbness and weakness in the arms, legs, or trunk; difficulty walking; and problems with bowel and bladder control.^{1,2}

Relieving symptoms will be a part of the treatment and care plan.

DIAGNOSIS

Doctors use different types of tests to detect a brain or spine tumor as well as determine its type and location.²⁰ These tests are often done by different specialists and providers who are part of the healthcare team.

DIFFERENT TESTS

After getting a thorough medical history from the patient (or caregiver in the case of a young child), the doctor will do a physical examination.³ A **neurological exam** tests the patient's eye movements, vision,

hearing, balance, coordination, and reflexes as well as cognitive skills such as awareness, attention, speech, language, memory, and judgment. This test is used to help determine which part of the brain or spine the tumor is affecting.

These tests will be done by either a *neurologist*, a physician trained in diagnosing and treating diseases and disorders of the CNS; a *neurosurgeon*, a physician trained in performing surgery for CNS tumors; an *oncologist or neurooncologist*, a doctor who specializes in treating patients with CNS tumors; or a *neuropsychologist*, a psychologist who specializes in understanding the relationship between the brain and behavior.

A **computed tomography (CT) scan**, a form of imaging, is typically ordered first as it is very helpful in identifying important features of the tumor, such as hydrocephalus.²

A **magnetic resonance imaging (MRI) scan** is the preferred test for further evaluating brain and spine tumors, such as ependymoma, because it shows a more detailed picture of the brain, spine, and tumor than a CT scan.^{2,10} These imaging tests can be used with or without contrast (gadolinium), which is a dye that makes the scanned images clearer.¹⁸ The doctor may want to do an **MRI of the spine** if the tumor is believed to be in the spine or to learn whether the ependymoma has spread from the brain to the CSF and spine.



A patient being prepped for a CT scan.

The doctor may recommend a **lumbar puncture**, also called a *spinal tap*, to look for tumor cells within the CSF.²⁰ A lumbar puncture is only done after the pressure in the brain or spine is relieved or the tumor is removed through surgery.

Although imaging tests may give the doctor an educated idea of the tumor type, a **biopsy** or surgical resection is needed to be sure of the diagnosis.²⁰ During a biopsy, the *neurosurgeon* surgically removes a small piece of the tumor tissue to send to a *pathologist* or *neuropathologist*, who is trained in identifying diseases of the CNS. The neuropathologist examines the sample under a microscope to determine the tumor type and grade. These results are included in the **pathology report** sent to the neurosurgeon and the rest of the healthcare team.

The neurosurgeon may remove a larger part of the tumor to send it for pathology testing. This is known as a **surgical resection**. A biopsy can be done as a separate procedure or as part of a surgical resection. The surgical resection can be complete (also called a **gross total resection**),¹⁸ if all visible tumor is safely removed, or incomplete (also called **partial or subtotal resection**), if it is not possible to remove the tumor completely in a safe way.

Not all these tests will be used for every patient. The doctor may consider the following factors when choosing which diagnostic tests to use:²⁰

- Type of tumor suspected
- Signs and symptoms
- Age and general health
- Results of other medical tests

Doctors are increasingly using **genetic tests** to determine the subtype of an ependymoma. Testing may involve a blood test in addition to genetic testing on

the tumor. If the tumor sample is sent to a laboratory that specializes in genetic testing, the doctor will get a report with the test results. Knowing the tumor subtype will help the doctor select the best treatment options.

TUMOR STAGING

When the diagnostic tests are complete, the doctor can stage the tumor. Staging is a way to describe where the tumor is located, if or where it has spread, and if it is affecting other parts of the CNS or body.^{3,23} Knowing the tumor stage will help determine the best treatment options and can even help predict a patient's prognosis.

Although there is no formal staging system for ependymoma, **the tumor can be classified based on where it is found** as follows:^{21,23}

- A supratentorial tumor grows above the cerebellum
- An infratentorial tumor grows in the cerebellum or brainstem
- A spinal tumor grows in the spinal cord

Some ependymomas are able to spread from their original location in one of these parts of the CNS to another.

Talk to your doctor about running genetic tests as they are not yet considered 'standard of care.'

TREATMENT

In general, **current treatment options** (often referred to as *standard of care*) for ependymoma, are surgery, radiation therapy, and in certain situations chemotherapy.¹

However, different subtypes may be treated differently.⁵ Many patients won't need additional treatment beyond surgery, depending on the tumor subtype and provided that the whole tumor is removed. For example, a

subependymoma can be cured by surgery alone and does not typically recur if it is removed completely.¹⁸ If a patient with ependymoma needs more treatment after surgery, it would be either radiation therapy or chemotherapy. However, the role of chemotherapy remains unproven and is still being studied.^{24,25} For now, chemotherapy is often limited to the treatment of very young children and some adults with a recurrent tumor.^{24,26}

Treatment options are based on:^{1,2,27}

- The tumor size, subtype, grade, and location
- What parts of the brain or spine the tumor is located in
- Current symptoms caused by the tumor
- If the tumor has invaded other parts of the brain or spine
- Possible side effects of treatment
- The patient's overall health, age, and preference

SURGERY

Having surgery to remove the tumor is an important first step in treating ependymoma.² The goals of surgery are to remove as much tumor as possible and confirm the diagnosis with a tissue sample for biopsy.^{2,25}

Research suggests that a patient's prognosis is better when all of the ependymoma visible to the neurosurgeon's eye and in the MRI is removed safely.^{2,6} In other words, when a **gross total resection** is done. While the goal is to fully remove the tumor, some ependymomas cannot be removed completely because of their location.²⁷ If that is the case, then a **subtotal** or **partial resection**¹⁸ will be performed to remove some of the tumor and get a tissue sample to confirm the diagnosis.

Thanks to recent medical advances, many tools are available to help the neurosurgeon surgically remove the tumor.²⁷ These include brain-mapping tools, special computer software, special dyes, and tiny instruments.

Common side effects of surgery include scalp pain,

swelling, scarring, and headaches.¹⁸ More severe side effects may include neurological or nerve damage, bleeding, infection, stroke, seizure, muscle paralysis, and wound healing problems.^{2,6}

A rare complication primarily in children with tumors located in the cerebellum is *cerebellar mutism syndrome*, also called *posterior fossa syndrome*.⁶ Symptoms include loss of speech and balance, muscle stiffness, fatigue, and personality changes. It is unclear why this syndrome occurs. For many children, the symptoms go away quickly, but others may have the symptoms years after surgery.¹⁰

Within days after surgery, a brain MRI will be done to look at how much tumor, if any, is remaining in the brain.¹⁸ For a spinal tumor, a spine MRI will be done after surgery for the same purpose. The amount of residual or remaining tumor will help the doctor make recommendations for further treatment.

Additional treatment will depend on:²⁷

- Amount of tumor removed
- Patient's age
- Whether the tumor has spread

In the case of a subtotal resection, the neurosurgeon may recommend a second surgery to try to remove the rest of the ependymoma.^{2,18} In children, **staged surgeries** are often performed.² The neurosurgeon will remove part of the tumor during the first surgery, followed by chemotherapy or radiation therapy to shrink the tumor. After several months, the neurosurgeon will do a second surgery to remove any residual tumor.

To learn more, read the ABTA's Surgery brochure.

It's important to weigh the benefits and risks of surgery as well as other treatment options.

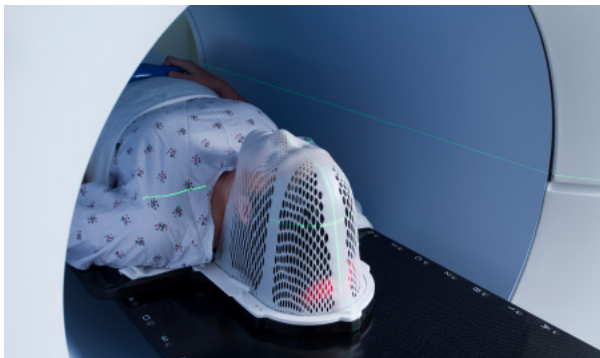
RADIATION

If the tumor cannot be removed surgically or only some of it is removed, the doctor may recommend radiation therapy to slow or stop the ependymoma from growing.²⁷ Radiation therapy uses very focused, high-energy rays to kill the tumor cells that remain in the brain.²⁸ A doctor who specializes in prescribing radiation therapy is called a *radiation oncologist*.

The most common type of radiation used to treat ependymomas is known as **external beam radiation therapy, or EBRT**.² There are different EBRT methods, but all of them involve using a machine to deliver the radiation through the skin directly to the tumor.²⁸ These include:

- Conventional radiation therapy
- Intensity modulated radiation therapy
- Proton therapy
- Stereotactic radiosurgery
- Three-dimensional conformal radiation therapy

All these techniques deliver a precise amount of radiation to the tumor and limit the amount of radiation to nearby healthy brain tissue.²¹ Reducing radiation exposure could result in fewer side effects due to the treatment.



Patient receiving targeted radiation therapy while wearing a customized mask for head stabilization during treatment.

Some doctors believe that all patients with low-grade (grades I and II) ependymoma should receive radiation no matter how much of the tumor is removed during surgery.² But, in general, recommendations for radiation therapy are based on several factors, including the type of surgery a patient has had and the tumor grade.⁶

For patients who have had a gross total resection of a grade I or grade II ependymoma in the brain that has not spread, National Comprehensive Cancer Network (NCCN) experts suggest that doctors consider doing EBRT.¹⁸ However, in the case of a supratentorial tumor that was removed completely, the patient can be carefully watched. This is known as *observation* and may be a good option depending on the specific tumor. Observation involves returning to the clinic and repeating the neurological examination and one or more tests, such as MRIs, over time to monitor the ependymoma's behavior. A major factor in choosing observation is determining whether the benefits of treatment outweigh the potential side effects. In children, all grade III ependymomas as well as all grade II and III ependymomas that occur near the cerebellum should receive EBRT.^{26,29}

Regarding spinal tumors, for patients who have had a gross total resection of a grade I or grade II ependymoma that has not spread, NCCN experts advise observation.¹⁸

In children with myxopapillary ependymoma, however, EBRT may be a better option than observation. That's because a myxopapillary ependymoma, although considered grade I with a favorable prognosis, may grow more rapidly in children and have a worse outcome.^{4,30} Studies suggest that radiation therapy following surgery for myxopapillary ependymoma can better control the disease and keep it from progressing. In adults with myxopapillary ependymoma, even if a complete resection was obtained, if the capsule of the tumor was disrupted, the chances of the tumor returning may be higher and irradiation may be recommended.³²

For both brain and spinal ependymomas, NCCN experts suggest performing EBRT for patients whose grade I or II tumor was not fully removed and for those who had a total resection of a grade III ependymoma that has not spread.¹⁸ If any of these tumors have spread, regardless of grade, radiation directed at the whole brain and spine, which is known as *craniospinal radiation therapy*, may be recommended.

There has been some concern about using radiation therapy for children younger than 3 years of age since irradiating the CNS can interfere with a child's growth and development. However, ongoing clinical trials suggest that radiation therapy is safe and effective even for very young children with ependymoma.^{8,21,27}

Generally, radiation therapy is given in a series of treatments over several weeks called *fractions*.^{2,28} Spacing the treatments out allows enough radiation to get into the body to kill the tumor, while giving the healthy cells time to recover.

Different people experience different side effects from radiation therapy. **Common side effects** include edema (swelling in the brain or spine near the tumor), fatigue (extreme tiredness even after sleeping), mild skin reactions, mouth irritation, hair loss, nausea, diarrhea, and cognitive and neurologic problems.^{2,27} Most of these side effects stop once treatment is finished. But some people also have long-term side effects that may include problems with growth, low hormone levels, hearing loss, and learning disabilities.⁶ **To learn more, read the ABTA's brochures on Conventional Radiation Therapy, Proton Therapy, and Stereotactic Radiosurgery.**

Talk to your doctor about the different types of radiation therapy and which one might be right for you or your child.

CHEMOTHERAPY

Chemotherapy uses powerful chemicals, or drugs, to stop or slow the growth of tumor cells.²⁷ It can be given through an intravenous (IV) tube into one's vein, usually in the arm, or a pill that is swallowed. **Chemotherapy is used to kill any remaining tumor cells** following surgery and/or radiation therapy or for tumors that return after the initial treatment and cannot be treated again with surgery or irradiation. It is prescribed by an *oncologist* or *neurooncologist*.



Patient receiving chemotherapy.

Chemotherapy drugs currently being used to treat ependymoma include cisplatin, carboplatin, cyclophosphamide, and etoposide, and temozolomide combined with lapatinib (a targeted therapy), among others.^{6,10,33} Often, more than one chemotherapy medication at a time is recommended because they attack the tumor in different ways and may be more effective when used together.^{6,27} Chemotherapy drugs used to treat CNS tumors are always being evaluated to develop new ones or new approaches to using them.²⁷

In general, the use of chemotherapy for the treatment of ependymoma is reserved for adults with recurrent tumor when further surgery and irradiation are no

longer options and for very young children in place of radiation therapy.^{6,24,26} If there is tumor left after chemotherapy, patients may undergo a second-look surgery to remove any remaining tumor.⁶ Similarly, older children who have had their tumors partially removed may benefit from chemotherapy, followed by a second-look surgery and irradiation. Researchers continue to study different ways to use chemotherapy before, during, or after radiation therapy.⁸

Conventional chemotherapy has been considered by many to be ineffective in treating ependymoma, but that opinion may be changing based on emerging research.

Chemotherapy is usually scheduled for a specific number of cycles given over a set period of time.²⁷

Common side effects of chemotherapy include fatigue, risk of infection, nausea and vomiting, hair loss, loss of appetite, and diarrhea. Side effects, which can depend on the dose given, usually stop after treatment is finished. **To learn more, read the ABTA's Chemotherapy brochure.**

PALLIATIVE CARE

Ependymoma and its treatments cause physical symptoms and side effects.²⁷ Relieving these symptoms and side effects is an important part of **supportive care**, sometimes referred to as *palliative care*, which is not the same as end-of-life care. Palliative care is for anyone, regardless of their age, or tumor type and stage. It should be started shortly after diagnosis for best results. **People who receive palliative care often have less severe symptoms, better quality of life, and are more satisfied with treatment.**²⁷

Palliative care may include medications, nutritional changes, relaxation techniques, and emotional and

spiritual support, among others.²⁷ As an example, the same drugs used to treat epilepsy may be prescribed to control seizures caused by ependymoma.¹⁰ These include antiepileptic and anticonvulsant drugs, such as levetiracetam, lacosamide, phenytoin, and carbamazepine. Steroids may be used to reduce the edema that is sometimes caused by the tumor or treatments and can cause pain or worsening of neurological symptoms. The most commonly used steroid is dexamethasone. Antiemetic medications prevent vomiting and help control nausea caused by the tumor or by irradiation or chemotherapy.

Using these drugs to treat symptoms is typically part of the care provided by the same healthcare team taking care of tumor treatment. However, sometimes a referral to a palliative care specialist is beneficial to control symptoms such as persistent pain or nausea.

Ask your doctor which therapies can be used to treat symptoms and side effects.

CLINICAL TRIALS

Clinical trials offer individuals **the chance to use new or experimental tests and treatments** (meaning they have not yet been proven) before they are available to the public. However, there is the risk that the treatment being studied may not work or comes with severe side effects.³⁴

Among the drugs being tested for the treatment of ependymoma in clinical trials are targeted therapies and immunotherapies.^{1,3}

Targeted therapies refer to medications that target certain proteins that help cause the ependymoma to grow and survive.³⁵ Unlike chemotherapy that can kill all

cells that are dividing rapidly, targeted therapy is more precise in killing specific tumor cells with a specific abnormal protein.

Immunotherapies refer to drugs that enlist the body's own immune system to fight the tumor.

Some clinical trials focus on new ways to relieve symptoms and side effects.⁸ Others focus on new medications and treatment combinations specifically for recurrent tumors.³⁵

People who want to join a clinical trial *volunteer* and must meet certain rules called *eligibility criteria*, such as having a specific type of tumor or not having been treated with a certain therapy. **To learn more, read the ABTA's Clinical Trials brochure.**

Talk to your doctor to see if a clinical trial is a good option for you or your child. More than 60% of children with cancer are treated in clinical trials.²⁷

FOLLOW-UP

Following treatment for ependymoma, **patients should meet routinely with their healthcare team to:**³⁶

- Manage ongoing side effects
- Check if the tumor has returned
- Monitor overall health

Follow-up visits may include a history and physical examination, an MRI of the brain and/or spine, and other medical tests.³⁶ MRIs are performed more often during the first few years after treatment and less often as times goes on.

Typically, patients will be seen every three to four months for the first three years, and then every four to six months for three to five years.¹⁸ However, more frequent visits may be required for patients with grade III tumors, who are currently undergoing irradiation or chemotherapy, or who recently completed these treatments. After five years, annual MRIs are typically recommended.²¹

Some patients may experience side effects from treatments long after the therapy has stopped.^{36,37}

These are called *long-term side effects*. Also, other side effects called *late effects* may start months or even years after active treatment is done. Late effects are not limited to the CNS. They may include heart and lung problems, growth and hormone issues, and the rare occurrence of a second type of cancer. Emotional problems, such as anxiety and depression, and a decline in cognitive skills have been known to occur as late effects.^{3,36}

Treating late effects may require seeing other specialists, such as a neurooncologist, neuropsychologist, or an *endocrinologist*, who is a doctor trained in treating growth or hormone imbalances. Some patients may benefit from rehabilitation, ranging from physical therapy, family or individual counseling, nutritional planning, and/or educational help.^{36,38}

Keeping a personal health record is important for patients with ependymoma, who should be monitored for the rest of their lives for recurrence and side effects.³⁶ Some patients will continue to see their oncologist, while others will go back to their family doctor. At some point in the person's life, it is likely that a doctor who was not directly involved in his or her initial care will be leading the patient's follow-up care.

When a patient no longer has any symptoms and the tumor cannot be found in the brain or spinal cord, it is referred to as being in *remission*.²⁷ Many remissions are

permanent, but it's still important to understand the risk of recurrence, that is, the chance of the tumor returning.

Talk to your doctor about the ongoing medical care that will be needed after treatment for ependymoma is finished.

RECURRENCE

When an ependymoma returns after treatment, it is called a recurrent tumor.²⁷ The recurring tumor can either come back in the same place it was first found or travel to other places in the brain or spine. About 80% of recurring ependymomas return to the same place.⁶ If a tumor returns, it usually does so within the first several years after the initial diagnosis, although it can occur 10 or even 20 years later.^{21,31,36}

When a tumor does return, the doctor will perform a new series of tests similar to those used to diagnose the original tumor to learn as much about the recurrent tumor as possible.²⁷ These tests may include blood tests, imaging scans, and a lumbar puncture. The doctor will recommend testing based on such factors as the type of tumor and stage originally diagnosed and the type of treatment given.³⁶

There is no standard treatment option for recurrent ependymoma.⁶ Treatment options should take into account the patient's age, location of the original and recurrent tumor, whether the ependymoma has spread, prior treatment, and the patient's ability to perform normal daily activities, sometimes referred to as *functional status*.

In general, treatments for a recurrent ependymoma may include:²⁷

- Additional surgery
- Radiation therapy
- Chemotherapy

These treatments may be given in a different combination or on a different schedule than was used with the first tumor. Having an additional surgery for recurrent tumors is associated with improved survival.^{6,26} Patients have had a positive response with repeat radiation therapy as well. Chemotherapy may be recommended for adults with recurrent tumor when further surgery and radiation therapy are no longer options.

Talk to your doctor about the possibility of the tumor recurring and treatment options available.

PROGNOSIS

Prognosis refers to the chance of recovery or survival from a disease.³ A prognosis is based on statistics that look at a large group of people with the same disease over time. Keep in mind that **statistics on survival rates are estimates and cannot accurately predict what is going to happen to an individual.** Typically, they are measured every five years, so the latest estimates may not include the most current methods of diagnosing and treating ependymoma.¹⁴

A patient's prognosis should be individualized and may take into account the following factors:

- **How much tumor remains in the brain or spine after surgery**
The prognosis is better when all the tumor can be surgically removed.^{4,6}

- **Whether the tumor has spread to other parts of the body**

While spreading to other parts of the body is linked with a poorer prognosis,³⁹ ependymomas tend not to spread outside the CNS.¹

- **The tumor's genetic subtype**

Subependymomas are associated with a very favorable prognosis when treated with gross total resection.^{2,6}

- **Age at diagnosis**

Generally, older children tend to have a better prognosis than younger children.^{6,21}

Recent research suggests that the amount of tumor removed is the most important predictor of survival.¹⁰ Survival rates for ependymomas are about 94% at one year, about 84% at five years, and about 79% at 10 years.^{11,12}

Most patients with a brain tumor can be treated successfully.²⁷ But when the tumor cannot be controlled, it is called *advanced* or *terminal ependymoma*.

Hospice care offers the best quality of life for people who are not expected to live longer than six months. Hospice care can be provided at home or a healthcare setting. In-home hospice care requires nursing care and special equipment, but it may allow some patients to continue engaging in social activities. For example, children may be happier if they can go to school, at least part time. The healthcare team can help determine what level of activity is acceptable to ensure that the patient has no pain or physical discomfort. Support services are available that can help individuals cope with an advanced tumor diagnosis.

Talk to your doctor about expected outcomes to get a more individualized prognosis.

FUTURE DIRECTIONS

Although research in the genetics of CNS tumors is very new, it has allowed researchers to further categorize ependymoma in ways that are having a significant impact on both treatment and survival. These genetic distinctions are important because they can help doctors better predict a patient's prognosis.⁸ Genetic testing has identified several genes associated with a person's risk of having a tumor return, the age at diagnosis, and tumor location.¹⁰

Through genetic testing, researchers are beginning to link tumor location with subtype.^{1,2} As an example, they have found that subependymomas tend to occur in the cerebellum while myxopapillary ependymomas tend to occur in the spine.

Researchers also have learned more about the incidence of ependymoma. They have found, for example, that classic ependymomas tend to occur more frequently in children whereas subependymomas occur more often in middle-aged and older people.⁴

The hope is that this growing knowledge will lead to better and more precise treatment based on tumor subtype.⁶ Knowing the genetic make-up of the tumor is an important first step in developing drugs that target and kill the cancer cells. Researchers continue to identify additional subtypes that will likely be added to the next WHO classification update.^{24,40}

Research efforts also are attempting to decrease the side effects of treatment therapies, while keeping high cure rates in patients with ependymoma. The goal is to improve survival and quality of life after therapy.

But there is more work to be done. Together, the medical and scientific communities, supporting organizations, and the patients and their families are building on past successes toward a better cure for all persons diagnosed with ependymoma.

AMERICAN BRAIN TUMOR ASSOCIATION INFORMATION, RESOURCES AND SUPPORT

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

GENERAL INFORMATION

About Brain Tumors: A Primer for Patients and Caregivers

Brain Tumor Dictionary*

Brain Tumors Handbook for the Newly Diagnosed*

Caregiver Handbook*

TUMOR TYPES

Ependymoma

Glioblastoma and Anaplastic Astrocytoma

Medulloblastoma

Meningioma

Metastatic Brain Tumors

Oligodendroglioma and Oligoastrocytoma

Pituitary Tumors

TREATMENT

Chemotherapy

Clinical Trials

Conventional Radiation Therapy

Proton Therapy

Stereotactic Radiosurgery*

Steroids

Surgery

AMERICAN BRAIN TUMOR ASSOCIATION INFORMATION, RESOURCES AND SUPPORT

INFORMATION

ABTA WEBSITE | ABTA.ORG

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

EDUCATION & SUPPORT

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

GET INVOLVED

- Join an ABTA fundraising event.
- Donate by visiting abta.org/donate.

CONTACT THE ABTA

CareLine: 800-886-ABTA (2282)

Email: abta cares@abta.org

Website: abta.org

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AMERICAN BRAIN TUMOR ASSOCIATION

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

For more information:

Website: abta.org

CareLine: 800-886-ABTA (2282)

Email: abtacares@abta.org



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