

Treating Brain and Spinal Cord Tumors in Children

If your child has been diagnosed with a brain or spinal cord tumor, your child's treatment team will discuss the options with you. It's important to weigh the benefits of each treatment option against the possible risks and side effects.

How are brain and spinal cord tumors treated?

The main treatments for children with brain and spinal cord tumors are:

- Surgery for Brain and Spinal Cord Tumors in Children
- Radiation Therapy for Brain and Spinal Cord Tumors in Children
- Chemotherapy for Brain and Spinal Cord Tumors in Children
- Targeted Therapy Drugs for Brain and Spinal Cord Tumors in Children
- Drugs to Help with Symptoms in Children with Brain or Spinal Cord Tumors

Common treatment approaches

In many cases children will get some combination of these treatments. Treatment is based on the type of tumor and other factors. Doctors plan each child's treatment individually to give them the best chance of a cure while limiting side effects as much as possible.

• Treating Specific Types of Childhood Brain and Spinal Cord Tumors

Who treats brain and spinal cord tumors in children?

Going through cancer treatment with a child often means meeting lots of specialists and

learning about parts of the medical system you probably haven't had contact with before.

Children and teens with brain and spinal cord tumors and their families have special needs that can be met best by cancer centers for children and teens, working closely with the child's primary care doctor. These centers offer the advantage of being treated by teams of specialists who know the differences between cancers in adults and those in children and teens, as well as the unique needs of younger people with cancer.

For childhood brain and spinal cord tumors, this team is often led by a **pediatric neurosurgeon**, a doctor who uses surgery to treat brain and nervous system tumors in children. Other doctors on the team may include:

- Pediatric neurologist: a doctor who treats brain and nervous system diseases in children
- Radiation oncologist: a doctor who uses radiation to treat cancer
- **Pediatric oncologist:** a doctor who uses chemotherapy and other medicines to treat children's cancers
- Endocrinologist: a doctor who treats diseases in glands that secrete hormones

Many other health professionals may be involved in your child's care as well, including physician assistants (PAs), nurse practitioners (NPs), nurses, psychologists, social workers, rehabilitation specialists, and other health professionals.

- How to Find the Best Cancer Treatment for Your Child
- Navigating the Health Care System When Your Child Has Cancer

Making treatment decisions

It's important to discuss your child's treatment options, including their goals and possible side effects, with the treatment team to help make the decision that's the best fit for your child. You may feel that you need to decide quickly, but it's important to give yourself time to absorb the information you have learned. It's also very important to ask questions if there is anything you're not sure about.

If time allows, getting a second opinion from another doctor experienced with your child's type of tumor is often a good idea. This can give you more information and help you feel more confident about the treatment plan you choose. If you aren't sure where to go for a second opinion, ask your doctor for help.

Your child might also see a psychologist or rehabilitation specialist before treatment begins. For example, if the tumor is slow growing and your child's condition is stable, they may be seen by a psychologist before treatment to assess any damage the tumor might have caused. Most of the work of these specialists takes place after treatment.

- Questions to Ask About Your Child's Brain or Spinal Cord Tumor
- How to Talk to Your Child's Cancer Care Team
- Seeking a Second Opinion

Thinking about taking part in a clinical trial

Today, most children and teens with cancer are treated at specialized children's cancer centers. These centers offer the most up-to-date-treatment by conducting clinical trials (studies of promising new therapies). Children's cancer centers often conduct many clinical trials at any one time, and in fact most children treated at these centers take part in a clinical trial as part of their treatment. Clinical trials are one way to get state-of-the art cancer treatment. Sometimes they may be the only way to get access to newer treatments (although there is no guarantee that newer treatments will be better). They are also the best way for doctors to learn better methods to treat these cancers. Still, they might not be right for everyone. If you would like to learn more about clinical trials that might be right for your child, start by asking the treatment team if your clinic or hospital conducts clinical trials.

<u>Clinical Trials</u>

Considering complementary and alternative methods

You may hear about alternative or complementary methods that your doctor hasn't mentioned to treat your child's tumor or relieve symptoms. These methods can include vitamins, herbs, and special diets, or other methods such as acupuncture or massage, to name a few. Complementary methods refer to treatments that are used along with your regular medical care. Alternative treatments are used instead of standard medical treatment. Although some of these methods might be helpful in relieving symptoms or helping people feel better, many have not been proven to work. Some might even be harmful. Be sure to talk to your child's cancer care team about any method you are thinking about using. They can help you learn what is known (or not known) about the method, which can help you make an informed decision.

<u>Complementary and Integrative Medicine</u>

Preparing for treatment

Before treatment, the doctors and other members of the team will help you, as a parent,

understand the tests that will need to be done. The team's social worker will also counsel you about the problems you and your child might have during and after treatments such as surgery, and might be able to help you find housing and financial aid if needed.

When Your Child Has Cancer

Help getting through cancer treatment

Your child's cancer care team will be your first source of information and support, but there are other resources for help when you need it. Hospital- or clinic-based support services can also be an important part of your care. These might include nursing or social work services, financial aid, nutritional advice, rehab, or spiritual help. For children and teens with cancer and their families, other specialists can be an important part of care as well. The American Cancer Society also has programs and services – including rides to treatment, lodging, and more – to help you get through treatment. Call our National Cancer Information Center at 1-800-227-2345 and speak with one of our trained specialists.

- Finding Help and Support When Your Child Has Cancer
- Programs & Services

The treatment information given here is not official policy of the American Cancer Society and is not intended as medical advice to replace the expertise and judgment of your cancer care team. It is intended to help you and your family make informed decisions, together with your doctor. Your doctor may have reasons for suggesting a treatment plan different from these general treatment options. Don't hesitate to ask your cancer care team any questions you may have about your treatment options.

Surgery for Brain and Spinal Cord Tumors in Children

- Surgery to remove or destroy the tumor
- Surgery to help with CSF flow blockage
- Surgery to place a ventricular access catheter
- Possible risks and side effects of surgery

More information about Surgery

For brain and spinal cord tumors, surgery may be done to:

- Get a <u>biopsy¹</u> sample to determine the <u>type of tumor</u>² and if the tumor cells have certain gene changes that might affect prognosis (outlook)
- Remove or destroy the tumor (or as much of it as possible)
- Help prevent or treat symptoms or possible complications from the tumor

Before surgery, be sure you understand what the goal of the surgery is, as well as the potential benefits and risks.

Surgery to remove or destroy the tumor

Most often, the first step in brain or spinal cord tumor treatment is for the neurosurgeon to remove or destroy as much of the tumor as possible, while trying to limit any effects on normal brain or nerve function.

Surgery alone or combined with radiation therapy may control or cure many slowergrowing tumors, including some low-grade astrocytomas, dysembryoplastic neuroepithelial tumors (DNETs), ependymomas, craniopharyngiomas, gangliogliomas, and meningiomas.

Children with tumors that tend to grow into nearby brain tissue, such as anaplastic astrocytomas or glioblastomas, cannot be cured by surgery alone. But surgery can reduce the amount of tumor that needs to be treated by radiation or chemotherapy, which might improve the results of these treatments.

Surgery can also help relieve some of the <u>symptoms</u>³ caused by brain tumors, particularly those caused by increased pressure within the skull, such as headaches, nausea, vomiting, and blurred vision. Surgery may also make seizures easier to control with medicines.

Surgery may not be a good option in some cases, such as if the tumor is deep within the brain, or if it has spread throughout a part of the brain that can't be removed, such as the brain stem. If this is the case, other treatments may be used instead.

Craniotomy

A craniotomy is a surgical opening made in the skull. This is the most common surgery

to remove a brain tumor. For this operation, the child may either be under general anesthesia (in a deep sleep) or may remain awake (with the surgical area numbed) for at least part of the surgery if brain function needs to be assessed during the operation.

Part of the head might need to be shaved before surgery. The neurosurgeon makes an incision (cut) in the skin over the skull near the tumor and then uses a special type of drill to remove a piece of bone from the skull. The opening is typically large enough for the surgeon to insert several instruments and view the parts of the brain needed to operate safely.

Many devices can help the surgeon see the tumor and surrounding brain tissue. The surgeon often operates while looking at the brain through a microscope. Imaging tests such as <u>MRI or CT scans</u>⁴ can be done before surgery (or ultrasound can be used once the skull has been opened) to help locate the tumor and its edges.

The surgeon will remove or destroy as much of the tumor as is safely possible. This can be done in several ways depending on how hard or soft the tumor is, and whether it has many or just a few blood vessels:

- One way is to cut it out with a scalpel or special scissors.
- Some tumors are soft and can be removed with suction devices.
- In other cases, a probe attached to an ultrasonic aspirator can be placed into the tumor to break it up and suck it out.

The surgeon is very careful to avoid damaging normal brain tissue as much as possible. To lower the risk of removing or damaging vital parts of the brain, different techniques can be used, such as:

- Functional MRI: Before surgery, this type of imaging test (described in <u>Tests for</u> <u>Brain and Spinal Cord Tumors in Children⁵</u>) can be done to locate a particular function of the brain. This can be used to help preserve that region during the operation.
- Intraoperative cortical stimulation (cortical mapping): During surgery, the surgeon can often detect the function of brain areas in and around the tumor by electrically stimulating them and monitoring the response. This will show if these areas control an important function, helping the surgeon to avoid them.
- Intraoperative imaging: In some cases, the surgeon uses MRI (or other) images taken at different times during the operation to show the location of any remaining tumor. This might allow some brain tumors to be removed more safely and extensively.

• Newer techniques: Newer types of MRI, as well as other techniques such as fluorescence-guided surgery, might be helpful in some situations. Some of these are described in <u>What's New in Research for Brain and Spinal Cord Tumors in</u> <u>Children?</u>⁶

After removing the tumor, the surgeon replaces the piece of skull bone and closes the incision. (If any metal screws, wires, or plates are needed to fasten the bone, they are usually made from titanium, which allows the child to get follow-up MRIs and will not set off metal detectors.)

For tumors that are hard to treat surgically, another option might be to insert a thin probe with a tiny laser on the end through a small hole in the skull and into the tumor. The laser is then used to heat and destroy (ablate) the tumor. This technique, known as **laser interstitial thermal therapy (LITT)**, is still fairly new, so doctors are still learning about the best ways to use it.

What to expect after surgery

After the operation to remove the tumor, the child may have a tube (called a *drain*) coming out of the incision that allows excess cerebrospinal fluid (CSF) to drain from the skull. Other tubes may be placed to allow blood that builds up after surgery to drain from under the scalp. The drains are usually removed after a few days. An imaging test such as an MRI or CT scan is typically done 1 to 3 days after the operation to confirm how much of the tumor has been removed. Recovery time in the hospital is usually 4 to 6 days, but this depends on the size and location of the tumor and whether other treatments are given.

Surgery to help with CSF flow blockage

If the tumor blocks the flow of CSF within the head, it can cause increased intracranial pressure (ICP) inside the skull. This can cause symptoms such as headaches, nausea, vomiting, and blurred vision, and may even damage the brain or be life-threatening. Surgery to remove the tumor can often help with this, but there are also other ways to drain away excess CSF and lower the pressure if needed.

Placing a shunt

The neurosurgeon may put in a silicone tube called a **shunt** (sometimes referred to as a ventriculoperitonealor**VP shunt**). One end of the shunt is placed in a ventricle of the brain (an area filled with CSF) and the other end is placed in the abdomen or, less

often, the heart (and would then be referred to as a *ventriculoatrial shunt*). The tube runs under the skin of the neck and chest, and allows the excess CSF to flow into the abdomen (or heart), where it mixes in with other fluids. The flow of CSF is controlled by a valve in the tubing.

Shunts can be temporary or permanent. They can be placed before or after the surgery to remove the tumor. Placing a shunt normally takes about an hour. Most children will need to stay in the hospital for about 1 to 3 days after the surgery. As with any operation, complications may develop, such as bleeding or infection. Sometimes shunts get clogged and need to be replaced.

Making an opening in the third ventricle

Another option to treat increased pressure in the skull in some cases is an **endoscopic third ventriculostomy**. In this operation, the surgeon makes an opening in the floor of the third ventricle at the base of the brain to allow the CSF to flow again. This operation is done through a small hole in the front of the skull. An advantage of this approach is that it does not require a shunt, but there is a chance that the opening made in the ventricle might close up again.

Placing an external drain

If the pressure inside the head needs to be relieved for a short time, an **external ventricular drain (EVD)** might be put in place to allow the excess CSF to drain out of the body. The drain is a small tube. One end is put into a ventricle, and the other end is attached to a collection bag outside the body. Along with collecting the excess CSF, the drain can also be used to measure the pressure inside the skull, as well as to look for tumor cells, blood, or signs of infection in the CSF.

The drain can be placed either during surgery or during a procedure at the hospital bedside. It can be put in place to relieve the pressure in the days before surgery, or to help drain the fluid that collects after an operation. If the pressure inside the skull needs to be lowered for more than a few days, the doctor might need to change this to a VP shunt.

Surgery to place a ventricular access catheter

Surgery may also be used to insert a **ventricular access catheter** to help deliver chemotherapy directly into the CSF later on. One type is called an Ommaya reservoir. A small incision is made in the scalp, and a small hole is drilled in the skull. A flexible tube is then inserted through the hole until the open end of the tube is in a ventricle, where it reaches the CSF. The other end, which has a dome-shaped reservoir, stays just under the scalp. After the operation, doctors and nurses can use a thin needle to give chemotherapy drugs through the reservoir or to remove CSF from the ventricle for testing.

Possible risks and side effects of surgery

Surgery on the brain or spinal cord is a serious operation, and surgeons are very careful to try to limit any problems either during or after surgery. Complications during or after surgery such as bleeding, infections, seizures, or reactions to anesthesia are rare, but they can happen.

Swelling in the brain is a major concern after surgery. Drugs called *corticosteroids* are typically given before and for several days after surgery to help lessen this risk.

One of the biggest concerns when removing brain tumors is the possible loss of brain function afterward, which is why doctors are very careful to remove only as much tissue as is necessary. Any symptoms of brain injury after surgery will depend mainly on the location and size of the tumor. If problems do arise, it might be right after surgery, or it might be days or even weeks later, so close monitoring for any changes is very important. (See <u>What Happens After Treatment for Brain and Spinal Cord Tumors in Children?</u>⁷)

More information about Surgery

For more general information about surgery as a treatment for cancer, see <u>Cancer</u> Surgery⁸.

To learn about some of the side effects listed here and how to manage them, see <u>Managing Cancer-related Side Effects</u>⁹.

Hyperlinks

- 1. <u>www.cancer.org/cancer/types/brain-spinal-cord-tumors-children/detection-</u> <u>diagnosis-staging/how-diagnosed.html</u>
- 2. <u>www.cancer.org/cancer/types/brain-spinal-cord-tumors-children/about/types-of-brain-and-spinal-tumors.html</u>

- 3. <u>www.cancer.org/cancer/types/brain-spinal-cord-tumors-children/detection-</u> <u>diagnosis-staging/signs-and-symptoms.html</u>
- 4. <u>www.cancer.org/cancer/types/brain-spinal-cord-tumors-children/detection-</u> <u>diagnosis-staging/how-diagnosed.html</u>
- 5. <u>www.cancer.org/cancer/types/brain-spinal-cord-tumors-children/detection-</u> <u>diagnosis-staging/how-diagnosed.html</u>
- 6. <u>www.cancer.org/cancer/types/brain-spinal-cord-tumors-children/about/new-research.html</u>
- 7. <u>www.cancer.org/cancer/types/brain-spinal-cord-tumors-children/after-</u> <u>treatment/follow-up.html</u>
- 8. <u>www.cancer.org/cancer/managing-cancer/treatment-types/surgery.html</u>
- 9. www.cancer.org/cancer/managing-cancer/side-effects.html

References

Chang SM, Mehta MP, Vogelbaum MA, Taylor MD, Ahluwalia MS. Chapter 97: Neoplasms of the central nervous system. In: DeVita VT, Lawrence TS, Rosenberg SA, eds. *DeVita, Hellman, and Rosenberg's Cancer: Principles and Practice of Oncology*. 10th ed. Philadelphia, Pa: Lippincott Williams & Wilkins; 2015.

Dorsey JF, Hollander AB, Alonso-Basanta M, et al. Chapter 66: Cancer of the central nervous system. In: Abeloff MD, Armitage JO, Niederhuber JE. Kastan MB, McKenna WG, eds. *Abeloff's Clinical Oncology*. 5th ed. Philadelphia, Pa: Elsevier; 2014.

Williams D, Parsons IF, Pollack DA. Chapter 26A: Gliomas, Ependymomas, and Other Nonembryonal Tumors of the Central Nervous System. In: Pizzo PA, Poplack DG, eds. *Principles and Practice of Pediatric Oncology*. 7th ed. Philadelphia, Pa: Lippincott Williams & Wilkins; 2016.

Last Revised: June 20, 2018

Radiation Therapy for Brain and Spinal

Cord Tumors in Children

- When might radiation therapy be used?
- Getting radiation therapy
- Special radiation therapy techniques
- Other types of radiation therapy
- Possible effects of radiation therapy
- Balancing the risks and benefits
- More information about radiation therapy

Radiation therapy uses high-energy x-rays or small particles to kill cancer cells. This type of treatment is given by a doctor called a **radiation oncologist**.

When might radiation therapy be used?

Radiation therapy may be used in different situations for brain or spinal cord tumors:

- · After surgery to try to kill any remaining tumor cells
- As part of the main treatment if surgery is not a good option
- To help prevent or relieve symptoms from the tumor

Children younger than 3 years are usually not given radiation because of possible longterm side effects with brain development. Instead, they are treated mainly with surgery and chemotherapy. Radiation can also cause some problems in older children. Radiation oncologists try very hard to deliver enough radiation to the tumor while limiting the radiation to normal surrounding brain areas as much as possible.

Getting radiation therapy

Most often, the radiation is focused on the tumor from a source outside the body. This is called **external beam radiation therapy (EBRT)**.

Before your child's treatments start, the radiation team will take careful measurements to determine the correct angles for aiming the radiation beams and the proper dose of radiation. This planning session, called *simulation*, usually includes getting imaging tests such as <u>CT or MRI scans</u>¹. Your child might be fitted with a plastic mold like a body cast to keep them in the same position so that the radiation can be aimed more accurately.

Most often, the total dose of radiation is divided into daily fractions (usually given Monday through Friday) over several weeks. For each treatment session, your child lies on a special table while a machine delivers the radiation from precise angles. Each treatment is much like getting an x-ray, but the dose of radiation is much higher. It is not painful. Some younger children might be given medicine to make them sleepy to make sure they don't move during the treatment. Each session lasts about 15 to 30 minutes, but most of the time is spent making sure the radiation is aimed correctly. The actual treatment time each day is much shorter.

Special radiation therapy techniques

Radiation therapy can damage normal brain tissue, so doctors try to deliver high doses of radiation to the tumor with the lowest possible dose to normal surrounding brain areas. Several techniques can help doctors focus the radiation more precisely:

Three-dimensional conformal radiation therapy (3D-CRT): 3D-CRT uses the results of imaging tests such as MRI and special computers to precisely map the location of the tumor. Several radiation beams are then shaped and aimed at the tumor from different directions. Each beam alone is fairly weak, which makes it less likely to damage normal tissues, but the beams join together at the tumor to give a higher dose of radiation there.

Intensity modulated radiation therapy (IMRT): IMRT is an advanced form of 3D therapy. In addition to shaping the beams and aiming them at the tumor from several angles, the intensity (strength) of the beams can be adjusted to limit the dose reaching the most sensitive normal tissues. This may let the doctor deliver a higher dose to the tumor. Many major hospitals and cancer centers now use IMRT.

Conformal proton beam radiation therapy: Proton beam therapy uses an approach similar to 3D-CRT. But instead of using x-rays, it focuses proton beams on the tumor. Unlike x-rays, which release energy both before and after they hit their target, protons cause little damage to tissues they pass through and then release their energy after traveling a certain distance. This means that more radiation can be delivered to the tumor, while doing less damage to the normal tissue around it. This approach may be more helpful for brain tumors that have distinct edges (such as chordomas), but it's not clear if it will be useful for tumors whose edges are mixed with normal brain tissue (such as astrocytomas or glioblastomas). There are only a limited number of proton beam centers in the United States at this time.

Stereotactic radiosurgery (SRS)/stereotactic radiotherapy (SRT): This type of treatment delivers a large, precise radiation dose to the tumor area in a single session

(SRS) or in a few sessions (SRT). It may be useful for some tumors in parts of the brain or spinal cord that can't be treated with surgery or when a child isn't healthy enough for surgery. (The term "radiosurgery" is used because the radiation is delivered so precisely, but there is no actual surgery involved in either SRS or SRT.)

For either procedure, a head frame is usually attached to the skull to help aim the radiation beams. Sometimes a face mask is used to hold the head in place instead. Once the exact location of the tumor is known from CT or MRI scans, radiation is focused at the tumor from many different angles. This can be done in 2 ways:

- In one approach, thin radiation beams are focused at the tumor from hundreds of different angles for a short period of time. Each beam alone is weak, but they all converge at the tumor to give a higher dose of radiation. The Gamma Knife is an example of a machine that uses this approach.
- Another approach uses a movable linear accelerator (a machine that creates radiation) that is controlled by a computer. Instead of delivering many beams at once, this machine moves around the head to deliver a thin beam of radiation to the tumor from many different angles. Several machines with names such as X-Knife, CyberKnife, and Clinac deliver stereotactic radiosurgery in this way.

SRS typically delivers the whole radiation dose in a single session, though it may be repeated if needed.

For SRT (also called *fractionated radiosurgery*) doctors give the radiation in several treatments to deliver the same or a slightly higher dose, which can now often be done without the need for a head frame.

Other types of radiation therapy

Brachytherapy (internal radiation therapy): Unlike the external radiation approaches above, in brachytherapy a radiation source is put directly into or near the tumor. The radiation it gives off travels a very short distance, so it affects only the tumor. This technique is most often used along with external radiation. It provides a high dose of radiation at the tumor site, while the external radiation treats nearby areas with a lower dose.

Whole brain and spinal cord radiation therapy (craniospinal radiation): If tests such as an MRI scan or lumbar puncture show the tumor has spread along the covering of the spinal cord (meninges) or into the surrounding cerebrospinal fluid, then external radiation may be given to the whole brain and spinal cord. Some tumors such as

ependymomas and medulloblastomas are more likely to spread this way, and therefore may require craniospinal radiation.

Possible effects of radiation therapy

Radiation is more harmful to tumor cells than it is to normal cells. Still, radiation can also damage normal brain tissue, especially in children younger than 3 years, which can lead to side effects.

Side effects during or soon after treatment: During radiation therapy, some children may become irritable and tired. Nausea, vomiting, and headaches are also possible but are uncommon. Spinal radiation causes nausea and vomiting more often than brain radiation. Sometimes dexamethasone (a corticosteroid) or other drugs can help relieve these symptoms. Some children might have hair loss in areas of the scalp that get radiation.

Some weeks after radiation therapy, children may become drowsy or have other nervous system symptoms. This is called the *radiation somnolence syndrome* or *early-delayed radiation effect*. It usually passes after a few weeks.

Problems with thinking and memory: Children may lose some brain function if large areas of the brain get radiation. Problems can include memory loss, personality changes, and trouble learning at school. These may get better over time, but <u>some effects may be long-lasting</u>².

Other side effects: Other effects could include seizures and slowed growth. There may also be other symptoms depending on the area of the brain treated and how much radiation was given.

Radiation necrosis: Rarely, a large mass of dead (necrotic) tissue forms at the site of the tumor in the months or years after radiation treatment. It can often be controlled with corticosteroid drugs, but surgery may be needed to remove the necrotic tissue in some instances.

Increased risk of another tumor: Radiation can damage genes in normal cells. As a result, there is a small risk of developing a second cancer in the area that got the radiation – for example, a meningioma of the coverings of the brain, another brain tumor, or less likely a bone cancer in the skull. If this occurs, it's usually many years after the radiation is given. This small risk should not keep children who need radiation from getting treatment. It's important to continue close follow-up with your child's doctor so that if problems do come up they can be found and treated as early as possible.

Balancing the risks and benefits

The risk of all of these side effects must be balanced against the risks of not using radiation and having less control of the tumor. If problems are seen after treatment, often it's hard to determine whether they were caused by damage from the tumor itself, from surgery or radiation therapy, or from some combination of these. Doctors are constantly testing lower doses or different ways of giving radiation to see if they can be as effective while causing fewer problems.

Normal brain cells grow quickly in the first few years of life, making them very sensitive to radiation. Because of this, radiation therapy is often not used or is postponed in children younger than 3 years old to avoid damage that might affect brain development. This needs to be balanced with the risk of tumor regrowth, because early radiation therapy may be lifesaving in some cases. It's important that you talk with your child's doctor about the risks and benefits of treatment.

More information about radiation therapy

To learn more about how radiation is used to treat cancer, see <u>Radiation Therapy</u>³.

To learn about some of the side effects listed here and how to manage them, see <u>Managing Cancer-related Side Effects</u>⁴.

Hyperlinks

- 1. <u>www.cancer.org/cancer/types/brain-spinal-cord-tumors-children/detection-</u> <u>diagnosis-staging/how-diagnosed.html</u>
- 2. <u>www.cancer.org/cancer/survivorship/children-with-cancer/late-effects-of-cancer-treatment.html</u>
- 3. www.cancer.org/cancer/managing-cancer/treatment-types/radiation.html
- 4. www.cancer.org/cancer/managing-cancer/side-effects.html

References

Chang SM, Mehta MP, Vogelbaum MA, Taylor MD, Ahluwalia MS. Chapter 97: Neoplasms of the central nervous system. In: DeVita VT, Lawrence TS, Rosenberg SA, eds. *DeVita, Hellman, and Rosenberg's Cancer: Principles and Practice of Oncology.* 10th ed. Philadelphia, Pa: Lippincott Williams & Wilkins; 2015. Dorsey JF, Hollander AB, Alonso-Basanta M, et al. Chapter 66: Cancer of the central nervous system. In: Abeloff MD, Armitage JO, Niederhuber JE. Kastan MB, McKenna WG, eds. *Abeloff's Clinical Oncology*. 5th ed. Philadelphia, Pa: Elsevier; 2014.

Williams D, Parsons IF, Pollack DA. Chapter 26A: Gliomas, Ependymomas, and Other Nonembryonal Tumors of the Central Nervous System. In: Pizzo PA, Poplack DG, eds. *Principles and Practice of Pediatric Oncology*. 7th ed. Philadelphia, Pa: Lippincott Williams & Wilkins; 2016.

Last Revised: June 20, 2018

Chemotherapy for Brain and Spinal Cord Tumors in Children

- When might chemotherapy be used?
- Which chemo drugs are used to treat brain and spinal cord tumors?
- Possible side effects of chemotherapy
- More information about chemotherapy

Chemotherapy (chemo) uses anti-cancer drugs that are usually given into a vein (IV) or taken by mouth. These drugs enter the bloodstream and reach almost all areas of the body. However, many chemo drugs aren't able to enter the brain and reach tumor cells.

For some brain tumors, the drugs can be given directly into the cerebrospinal fluid (CSF, the fluid that bathes the brain and spinal cord), either in the brain or into the spinal canal below the spinal cord. To help with this, a thin tube known as a *ventricular access catheter* may be inserted through a small hole in the skull and into a ventricle during a minor operation. (See Surgery for Brain and Spinal Cord Tumors in Children.)

When might chemotherapy be used?

In general, chemo is used for faster growing tumors. Some types of brain tumors, such as medulloblastomas, tend to respond well to chemo.

Chemo is most often used along with other types of treatment such as surgery and radiation therapy. It may be used instead of radiation therapy in children 3 years and

younger.

Which chemo drugs are used to treat brain and spinal cord tumors?

Some of the chemo drugs used to treat children with brain or spinal cord tumors include:

- Carboplatin
- Carmustine (BCNU)
- Cisplatin
- Cyclophosphamide
- Etoposide
- Lomustine (CCNU)
- Methotrexate
- Temozolomide
- Thiotepa
- Vincristine

These drugs may be used alone or in various combinations, depending on the type of brain tumor. Doctors give chemo in cycles. Each cycle generally lasts for a few weeks and is followed by a rest period to give the body time to recover.

Possible side effects of chemotherapy

Chemo drugs can cause side effects. These depend on the type and dose of drugs, and how long treatment lasts. Possible side effects can include:

- Hair loss
- Mouth sores
- Loss of appetite
- Nausea and vomiting
- Diarrhea
- Increased chance of infections (from having too few white blood cells)
- Easy bruising or bleeding (from having too few blood platelets)
- Fatigue (from having too few red blood cells or other factors)

Some of the most effective drugs against brain tumors tend to have fewer of these side effects than other common chemo drugs, but they can still occur. Most side effects tend to go away once treatment is finished. Your child's doctor and treating team will watch

closely for any side effects. There are often ways to lessen these side effects. For example, drugs can be given to help prevent or reduce nausea and vomiting.

Some chemo drugs can also have other, less common side effects. For example, cisplatin and carboplatin can also cause kidney damage and hearing loss. Your child's kidney function and hearing will be checked periodically if they are given these drugs.

Be sure to ask your child's doctor or nurse about medicines to help reduce side effects, and let them know if your child has side effects so they can be managed. In some cases, the doses of the chemo drugs may need to be reduced or treatment may need to be delayed or stopped to prevent the effects from getting worse.

More information about chemotherapy

For more general information about how chemotherapy is used to treat cancer, see <u>Chemotherapy</u>¹.

To learn about some of the side effects listed here and how to manage them, see <u>Managing Cancer-related Side Effects</u>².

Hyperlinks

- 1. www.cancer.org/cancer/managing-cancer/treatment-types/chemotherapy.html
- 2. <u>www.cancer.org/cancer/managing-cancer/side-effects.html</u>

References

Chang SM, Mehta MP, Vogelbaum MA, Taylor MD, Ahluwalia MS. Chapter 97: Neoplasms of the central nervous system. In: DeVita VT, Lawrence TS, Rosenberg SA, eds. *DeVita, Hellman, and Rosenberg's Cancer: Principles and Practice of Oncology*. 10th ed. Philadelphia, Pa: Lippincott Williams & Wilkins; 2015.

Dorsey JF, Hollander AB, Alonso-Basanta M, et al. Chapter 66: Cancer of the central nervous system. In: Abeloff MD, Armitage JO, Niederhuber JE. Kastan MB, McKenna WG, eds. *Abeloff's Clinical Oncology*. 5th ed. Philadelphia, Pa: Elsevier; 2014.

Williams D, Parsons IF, Pollack DA. Chapter 26A: Gliomas, Ependymomas, and Other Nonembryonal Tumors of the Central Nervous System. In: Pizzo PA, Poplack DG, eds. *Principles and Practice of Pediatric Oncology*. 7th ed. Philadelphia, Pa: Lippincott

Williams & Wilkins; 2016.

Last Revised: June 20, 2018

Targeted Therapy Drugs for Brain and Spinal Cord Tumors in Children

As researchers have learned more about the changes in the inner workings of cells that cause cancer or help cancer cells grow, they have developed newer drugs that target these changes. These targeted drugs work differently from standard <u>chemotherapy</u>¹ drugs. They sometimes work when chemo drugs don't, and they often have different side effects.

Targeted drugs do not yet play a large role in treating brain or spinal cord tumors, but some of them might be helpful for certain types of tumors.

- Drugs that target tumors with BRAF gene changes
- Drugs that target tumors with IDH gene changes
- Everolimus (Afinitor)
- More information about targeted therapy

Drugs that target tumors with BRAF gene changes

In some low-grade gliomas, the tumor cells have changes (mutations) in the *BRAF* gene, which causes them to make certain proteins that help the cells grow.

Dabrafenib (Tafinlar) and trametinib (Mekinist)

Dabrafenib and trametinib are targeted drugs that affect some of these proteins. (Dabrafenib targets the BRAF protein, while trametinib affects the related MEK protein.) Combining these drugs might be an option for children at least one year of age with any type of <u>low-grade glioma</u>² with a *BRAF* V600E mutation that needs treatment other than surgery or radiation. If your child has a low-grade glioma, the tumor cells might be <u>tested</u>³ for this gene change to see if these drugs might be helpful.

These drugs are typically taken daily, either as pills or as liquids.

Common **side effects** can include skin changes, rash, itching, sensitivity to the sun, headache, fever, chills, joint or muscle pain, fatigue, cough, hair loss, nausea, diarrhea, and high blood pressure.

Less common but serious side effects can include bleeding, heart rhythm problems, liver or kidney problems, lung problems, severe allergic reactions, severe skin or eye problems, and increased blood sugar levels.

Some people treated with these drugs have developed skin cancers, especially squamous cell skin cancers. Your child's doctor will want to check their skin often during treatment. You should also let the doctor know right away if you notice any new growths or abnormal areas on your child's skin.

Tovorafenib (Ojemda)

Tovorafenib targets the BRAF protein. This drug might be an option for children at least 6 months of age with any type of <u>low-grade glioma</u>⁴ that has a *BRAF* gene change and that has come back or is no longer responding to other treatments. If your child has a low-grade glioma, the tumor cells might be <u>tested</u>⁵ for BRAF gene changes to see if this drug might be helpful.

This drug is typically taken once a week, either as pills or as a liquid.

Common **side effects of tovorafenib** can include skin changes (such as rash, dry skin, or acne), sensitivity to the sun, hair color changes, feeling tired, headache, nausea and vomiting, fever, constipation, an increased risk of infections, and changes in some blood test results. This drug might also slow a child's growth.

Less common but serious side effects can include serious bleeding, liver problems, and severe reactions to sunlight.

Drugs that target tumors with *IDH* gene changes

In children with some types of brain tumors, the tumor cells might have a change (mutation) in either the *IDH1* or *IDH2* gene. These genes help the cells make certain proteins, which are also called IDH1 and IDH2. Mutations in one of these genes can stop the tumor cells from maturing the way they normally would.

Vorasidenib (Voranigo)

Vorasidenib is a type of targeted drug known as an **IDH inhibitor**. It blocks abnormal

IDH1 and IDH2 proteins, which seems to help the tumor cells mature into more normal cells.

This drug can be used after surgery in children 12 years of age and older with a <u>grade 2</u> <u>astrocytoma or oligodendroglioma⁶</u>, if the tumor cells are found to have an *IDH1* or *IDH2* gene mutation. The doctor can get tests of the tumor cells to see if they have one of these mutations.

This drug is taken by mouth as tablets, once a day.

Common **side effects of vorasidenib** can include feeling very tired, headache, nausea, muscle aches or stiffness, diarrhea, seizures, and changes in lab tests showing the drug is affecting the liver.

Sometimes this drug might have more serious effects on the liver, which could lead to symptoms such as jaundice (yellowing of the eyes and skin), dark urine, loss of appetite, or pain in the upper right side of the belly. It's important to let the health care team know if any of these symptoms appear.

Everolimus (Afinitor)

For subependymal giant cell astrocytomas (SEGAs) that can't be removed completely by surgery, everolimus may shrink the tumor or slow its growth for some time. This drug works by blocking a cell protein known as mTOR, which normally helps cells grow and divide into new cells.

Everolimus is a pill taken once a day.

Common **side effects of everolimus** can include mouth sores, increased risk of infections, nausea, loss of appetite, diarrhea, skin rash, feeling tired or weak, fluid buildup (usually in the legs), and increases in blood sugar and cholesterol levels. A less common but serious side effect is lung damage, which can cause shortness of breath or other problems.

Many other targeted drugs are now being developed and studied in clinical trials. Some of these are described in <u>What's New in Research and Treatment for Brain and Spinal</u> <u>Cord Tumors in Children?</u>⁷

More information about targeted therapy

To learn more about how targeted drugs are used to treat cancer, see Targeted Cancer

Therapy⁸.

To learn about some of the side effects listed here and how to manage them, see <u>Managing Cancer-related Side Effects</u>⁹.

Hyperlinks

- 1. <u>www.cancer.org/cancer/managing-cancer/treatment-types/chemotherapy.html</u>
- 2. <u>www.cancer.org/cancer/types/brain-spinal-cord-tumors-children/about/types-of-brain-and-spinal-tumors.html</u>
- 3. www.cancer.org/cancer/types/brain-spinal-cord-tumors-children/detectiondiagnosis-staging/how-diagnosed.html
- 4. <u>www.cancer.org/cancer/types/brain-spinal-cord-tumors-children/about/types-of-brain-and-spinal-tumors.html</u>
- 5. <u>www.cancer.org/cancer/types/brain-spinal-cord-tumors-children/detection-</u> <u>diagnosis-staging/how-diagnosed.html</u>
- 6. <u>www.cancer.org/cancer/types/brain-spinal-cord-tumors-children/about/types-of-brain-and-spinal-tumors.html</u>
- 7. <u>www.cancer.org/cancer/types/brain-spinal-cord-tumors-children/about/new-</u>research.html
- 8. www.cancer.org/cancer/managing-cancer/treatment-types/targeted-therapy.html
- 9. <u>www.cancer.org/cancer/managing-cancer/side-effects.html</u>

References

Chang SM, Mehta MP, Vogelbaum MA, Taylor MD, Ahluwalia MS. Chapter 97: Neoplasms of the central nervous system. In: DeVita VT, Lawrence TS, Rosenberg SA, eds. *DeVita, Hellman, and Rosenberg's Cancer: Principles and Practice of Oncology*. 10th ed. Philadelphia, Pa: Lippincott Williams & Wilkins; 2015.

Dorsey JF, Hollander AB, Alonso-Basanta M, et al. Chapter 66: Cancer of the central nervous system. In: Abeloff MD, Armitage JO, Niederhuber JE. Kastan MB, McKenna WG, eds. *Abeloff's Clinical Oncology*. 5th ed. Philadelphia, Pa: Elsevier; 2014.

Williams D, Parsons IF, Pollack DA. Chapter 26A: Gliomas, Ependymomas, and Other Nonembryonal Tumors of the Central Nervous System. In: Pizzo PA, Poplack DG, eds. *Principles and Practice of Pediatric Oncology*. 7th ed. Philadelphia, Pa: Lippincott Williams & Wilkins; 2016.

Last Revised: August 8, 2024

Drugs to Help with Symptoms in Children with Brain or Spinal Cord Tumors

Children with brain or spinal cord tumors can often be given drugs to help with symptoms from the tumor or side effects of treatment. These drugs do not treat the tumor directly, but they can be important an important part of your child's treatment.

Corticosteroids

Cortisone-like drugs such as dexamethasone (Decadron) are often given to reduce the swelling that can occur around brain tumors. They are often given before and for a few days after surgery, and might be used during radiation therapy as well. This may help relieve side effects such as headaches, nausea, and vomiting.

Anti-seizure drugs (anticonvulsants)

Drugs may be given to lower the chance of seizures in children with brain tumors. Many different anti-seizure drugs can be used.

Hormones

The pituitary gland, which lies at the base of the brain, helps control the levels of many different hormones in the body. If the pituitary has been damaged by the spread of the tumor or by treatments (surgery or radiation therapy), your child may need to take pituitary hormones or other hormones to replace those that are no longer being made by the body.

References

Chang SM, Mehta MP, Vogelbaum MA, Taylor MD, Ahluwalia MS. Chapter 97: Neoplasms of the central nervous system. In: DeVita VT, Lawrence TS, Rosenberg SA, eds. *DeVita, Hellman, and Rosenberg's Cancer: Principles and Practice of Oncology.* 10th ed. Philadelphia, Pa: Lippincott Williams & Wilkins; 2015.

Dorsey JF, Hollander AB, Alonso-Basanta M, et al. Chapter 66: Cancer of the central nervous system. In: Abeloff MD, Armitage JO, Niederhuber JE. Kastan MB, McKenna

WG, eds. Abeloff's Clinical Oncology. 5th ed. Philadelphia, Pa: Elsevier; 2014.

Williams D, Parsons IF, Pollack DA. Chapter 26A: Gliomas, Ependymomas, and Other Nonembryonal Tumors of the Central Nervous System. In: Pizzo PA, Poplack DG, eds. *Principles and Practice of Pediatric Oncology*. 7th ed. Philadelphia, Pa: Lippincott Williams & Wilkins; 2016.

Last Revised: June 20, 2018

Treating Specific Types of Childhood Brain and Spinal Cord Tumors

The treatment options for brain and spinal cord tumors in children depend on many factors, including:

- The type of tumor¹
- The location of the tumor
- How far the tumor has grown or spread
- The child's age and overall health
- Non-infiltrating (grade I) astrocytomas (pilocytic astrocytomas and subependymal giant cell astrocytomas)
- Diffuse astrocytomas (grade II)
- Higher-grade (grade III or IV) astrocytomas (anaplastic astrocytomas and glioblastomas)
- Oligodendrogliomas
- Ependymomas and anaplastic ependymomas
- Optic gliomas
- Brain stem gliomas
- *Targeted drugs for low-grade gliomas
- Embryonal tumors (including medulloblastomas)
- Meningiomas
- Schwannomas (including acoustic neuromas)
- Spinal cord tumors
- Choroid plexus tumors

- Craniopharyngiomas
- Germ cell tumors

Non-infiltrating (grade I) astrocytomas (pilocytic astrocytomas and subependymal giant cell astrocytomas)

Many doctors consider these to be benign tumors because they tend to grow very slowly and do not grow into (infiltrate) nearby tissues. **Pilocytic astrocytomas** occur most often in the cerebellum in young children, while **subependymal giant cell astrocytomas (SEGAs)** grow in the ventricles and are almost always seen in children with <u>tuberous sclerosis</u>².

Most children with these astrocytomas can be cured by **surgery** alone. They may be given radiation therapy if the tumor is not removed completely, although many doctors will wait until there are signs the tumor is growing back before considering it. Even then, another operation to remove the remaining tumor may be the first option. The outlook is not as good if the tumor is in a place that does not allow it to be removed surgically, such as the hypothalamus or brain stem. In these cases, radiation therapy is usually the best option.

For SEGAs that can't be removed completely by surgery, treatment with the targeted drug everolimus (Afinitor) might shrink the tumor or slow its growth for some time.

Other targeted drugs might also be an option for some of these tumors.*

Diffuse astrocytomas (grade II)

These tumors tend to grow slowly, but they can grow into nearby tissues. The initial treatment for these tumors is surgery if it can be done, or biopsy to confirm the diagnosis³ if surgery is not feasible. Because these tumors often grow into nearby normal brain tissue, they are hard to cure with just surgery. Usually the surgeon will try to remove as much of the tumor as safely possible. If the surgeon can remove it all, the child may be cured with no further treatment.

Radiation therapy may be given after surgery, especially if a lot of tumor remains. Otherwise, radiation may be postponed until the tumor starts to regrow. (Sometimes, a second surgery might be tried before giving radiation.) Radiation may also be used as the main treatment if surgery is not a good option because of the tumor's location.

For children younger than 3, if the tumor can't be removed completely or if it grows

back, chemotherapy may be used to try to slow the tumor's growth until they are older. They may then be treated with radiation.

Targeted drugs might also be an option for some of these tumors.*

Higher-grade (grade III or IV) astrocytomas (anaplastic astrocytomas and glioblastomas)

Surgery is often the first treatment for these fast-growing astrocytomas if it can be done, but patients with these tumors are almost never cured by surgery. In some cases only a <u>needle biopsy</u>⁴ is safe. When surgery is used, as much of the tumor is removed as possible, and then radiation therapy is given, often followed by chemotherapy.

If surgery can't be done, radiation is the main treatment, again often followed by chemotherapy.

If the child is younger than 3, radiation may be postponed until they are older. Surgery may be repeated in some cases if the tumor comes back after the initial treatment.

Because these tumors are hard to cure with current treatments, <u>clinical trials</u>⁵ of promising new treatments may be a good option.

Oligodendrogliomas

If possible, surgery is the first option for oligodendrogliomas. Although surgery does not usually cure these infiltrating tumors, it can relieve symptoms and prolong survival. Many of these tumors grow slowly, and surgery may be repeated if it grows back in the same spot. Radiation therapy and/or chemotherapy may be given after surgery.

If surgery is not an option, chemotherapy, with or without radiation therapy, may be helpful. Oligodendrogliomas may respond to chemotherapy better than other brain tumors if the tumor cells have certain chromosome changes. Ask your child's doctor about testing for these changes.

Targeted drugs might also be an option for some of these tumors.*

Ependymomas and anaplastic ependymomas

These tumors usually do not grow into nearby normal brain tissue. They can sometimes be cured by surgery if the entire tumor can be removed, but this isn't always possible. If

some of the tumor is left behind, a second operation may be done in some cases (often after a short course of chemotherapy).

Radiation therapy is recommended after surgery for most patients to try to prevent the tumor from coming back, even if it appears that all of the tumor has been removed.

The use of chemotherapy after surgery is still being tested in <u>clinical trials</u>⁶. Some doctors might recommend it, but its benefit is still uncertain. It may be more helpful for anaplastic ependymomas. Very young children may be given chemotherapy after surgery to help avoid or delay the use of radiation.

Targeted drugs might also be an option for some of these tumors.*

Sometimes ependymoma tumor cells can spread into the cerebrospinal fluid (CSF). A few weeks after surgery, the doctor may order an MRI scan of the brain and spinal cord and test the CSF for tumor cells by doing a lumbar puncture (spinal tap). If tumor cells are found in the CSF or growing on the surface of the nervous system, radiation is typically given to the entire brain and spinal cord.

Optic gliomas

These tumors start in the optic nerves (the nerves leading from the eyes to the brain). They are often hard to operate on because these nerves are very sensitive and might be harmed by surgery. Depending on where the tumor is, removing it could lead to loss of vision in one or both eyes, so the benefits and risks of surgery have to be considered carefully. In some cases, a child might have already lost some vision because of the tumor itself. Sometimes surgery might not be needed, because these tumors can grow very slowly.

If treatment is needed and the tumor can be removed completely, surgery is often the preferred treatment. But in many cases (especially if the child has <u>neurofibromatosis</u> type 1^7) the tumor is likely to have spread too far along the optic nerves to be removed completely. Radiation therapy is preferred for these tumors if treatment is needed, although it can also affect a child's vision (and can have other long-term side effects).

Younger children may get chemotherapy instead of radiation. Radiation could then be used later if needed as the child gets older.

Targeted drugs might also be an option for some of these tumors.*

Brain stem gliomas

Most of these tumors are astrocytomas, although a small number are ependymomas or other tumors. These tumors usually look a certain way on <u>MRI scans</u>⁸, so the diagnosis can often be made without surgery or a biopsy.

Focal brain stem gliomas: A small number of brain stem gliomas are small tumors with very distinct edges (called *focal* brain stem gliomas). Some of these tumors grow so slowly that treatment might not be needed unless the tumor causes problems. If treatment is needed, these tumors can often be treated successfully with surgery. If surgery can't be done, radiation therapy may be used to slow its growth. Radiation can also be used if surgery doesn't remove the tumor completely.

Diffuse brain stem gliomas: Most brain stem gliomas grow diffusely throughout the brain stem, rather than as a distinct (focal) tumor. These tumors often start in the pons, where they are called *diffuse intrinsic pontine gliomas* (DIPGs). The brain stem is vital to life and can't be removed, so surgery in these cases would most likely do more harm than good, and it is usually not attempted. Diffuse brain stem gliomas typically are treated with radiation therapy. Chemotherapy is sometimes added, although it's not clear if it's helpful.

Diffuse brain stem tumors are very hard to control. But in children with <u>neurofibromatosis type 1⁹</u>, these tumors often grow slowly (or even stop growing), so these children tend to have a better outlook. Because these tumors are hard to treat, <u>clinical trials¹⁰</u> of promising new treatments may be a good option.

Targeted drugs might also be an option for some brain stem gliomas if they are low grade.*

*Targeted drugs for low-grade gliomas

For any type of <u>low-grade (grade I or II) glioma¹¹</u>, the tumor cells might be tested for *BRAF* gene mutations. If the cells have a *BRAF* gene change, treatment with targeted drugs such as dabrafenib plus trametinib, or tovorafenib, might be an option at some point.

For grade II (2) astrocytomas or oligodendrogliomas, the tumor cells might be tested for *IDH1* and *IDH2* gene mutations. If the cells have one of these gene changes, a targeted drug such as vorasidenib might be an option after surgery.

Embryonal tumors (including medulloblastomas)

Embryonal tumors tend to grow quickly and spread through the spread to cerebrospinal

fluid (CSF). In the past, many embryonal tumors were referred to as **primitive neuroectodermal tumors (PNETs)**. Embryonal tumors are all treated in similar ways, but medulloblastomas tend to have a better outlook than other types.

Medulloblastomas: These tumors start in the cerebellum. They tend to grow quickly and are among those most likely to spread outside the brain (usually to the bones or the bone marrow). But they also tend to respond well to treatment.

These tumors can often block the flow of CSF. This can lead to a dangerous rise in the pressure inside the skull, which might need to be treated right away with some type of surgery.

Children with medulloblastoma are often divided into <u>standard-risk and high-risk</u> <u>groups</u>¹², depending on certain factors. Those in the high-risk group usually get more intensive treatment than children in the standard-risk group. More recently, doctors have started to divide these tumors into 4 groups based on the gene changes within the tumor cells. These might also be used to help determine the best treatment.

Medulloblastomas are removed with surgery when possible, followed by radiation therapy to the area where they started. High doses of radiation are aimed at the area of the tumor. Because these tumors tend to spread to the CSF, children 3 or older also may be given lower doses of radiation to the whole brain and the spinal cord (craniospinal radiation). Chemotherapy is usually given after radiation therapy, which might let doctors use lower doses of radiation in some cases. But if the tumor has spread through the CSF, standard doses of radiation will be needed.

For children younger than 3, doctors try to use as little radiation as possible. Chemotherapy is typically the first treatment given after surgery. Depending on how the tumor responds, chemotherapy might be followed by radiation therapy.

There are some reports that giving high-dose chemotherapy followed by an autologous stem cell transplant may be helpful for some children with medulloblastomas. Several clinical trials are now studying this. For more information on stem cell transplants, see <u>Stem Cell Transplant for Cancer¹³</u>.

Other embryonal tumors and pineoblastomas: Less common types of embryonal tumors include:

- Medulloepithelioma
- Atypical teratoid/rhabdoid tumor (ATRT)
- Embryonal tumor with multilayered rosettes

Pineoblastomas are no longer considered a type of embryonal tumor, but they are treated in a similar way.

These tumors also tend to grow quickly, and they are generally harder to treat than medulloblastomas (although treatment is often like that used for high-risk medulloblastomas).

Surgery is the main treatment for these tumors, but they usually are hard to remove completely. Still, surgery can relieve symptoms and may help make other treatments more effective. Children 3 or older are given radiation therapy after surgery. Because these tumors tend to spread to the CSF, radiation therapy is often given to the whole brain and the spinal cord (craniospinal radiation).

Chemotherapy may be given with radiation therapy so that a lower dose of radiation can be used. But if the tumor has spread to the CSF, standard doses of radiation will be required. Chemotherapy is also used to treat tumors that come back (recur).

For children younger than 3 years, doctors try to use as little radiation as possible. Chemotherapy is typically the first treatment given after surgery. Some studies have shown very good results using chemotherapy in young children. Depending on how the tumor responds, chemotherapy may or may not be followed by radiation therapy.

There are some reports that giving high-dose chemotherapy followed by an autologous stem cell transplant may be helpful for children with pineoblastomas and other types of embryonal tumors. Several clinical trials are now studying this. For more information on stem cell transplants, see <u>Stem Cell Transplant for Cancer¹⁴</u>.

Meningiomas

Surgery is the main treatment for these tumors. Children are usually cured if the surgery removes the tumor completely.

Some tumors, particularly those at the base of the brain, can't be removed completely, and some are invasive and come back even though they were thought to be completely removed. Radiation therapy after surgery may control the growth of these tumors. Chemotherapy may be tried if surgery and radiation aren't effective, but it is not helpful in many cases.

Schwannomas (including acoustic neuromas)

These slow-growing tumors are usually benign and are cured by surgery. In some centers, small vestibular schwannomas (also known as *acoustic neuromas*) are treated by stereotactic radiosurgery. For larger schwannomas where complete removal is likely to cause problems, as much as possible is safely removed, and what's left is treated with radiosurgery.

Spinal cord tumors

These tumors are usually treated similarly to those of the same type in the brain.

Astrocytomas of the spinal cord usually can't be removed completely. They may be treated with surgery to remove as much tumor as possible, followed by radiation therapy, or with radiation therapy alone. Chemotherapy may be used after surgery instead of radiation in younger children. It may also be used after radiation therapy in older children if the tumor appears to be growing quickly.

Meningiomas near the spinal cord are often cured by surgery. Some ependymomas can be cured by surgery as well. If an ependymoma can't be removed completely, radiation therapy will be given after surgery.

Choroid plexus tumors

Benign choroid plexus papillomas are usually cured with just surgery. Choroid plexus carcinomas are malignant tumors that are only sometimes cured by surgery. After surgery, these carcinomas are usually treated with radiation therapy and/or chemotherapy.

Craniopharyngiomas

Craniopharyngiomas grow very close to the pituitary gland, the optic nerves, and blood vessels that supply the brain, so they can be hard to remove completely without causing side effects. Some neurosurgeons prefer surgery to remove as much of the tumor as possible, while others prefer to remove most of the tumor (debulking) and then give radiation therapy.

Partial surgical removal followed by very focused radiation therapy may cause fewer severe side effects than complete removal, but it is not yet clear if this approach is as good at preventing the tumor from growing back.

Germ cell tumors

The most common germ cell tumor, **germinoma**, can usually be cured by radiation therapy alone (after it is diagnosed by surgery or looking at a cerebrospinal fluid sample). Chemotherapy may be added if the tumor is very large or if radiation doesn't destroy it completely. To try to reduce side effects in children who have not yet reached puberty, some doctors use chemotherapy followed by a reduced dose of radiation as the main treatment. In very young children, chemotherapy may be used instead of radiation therapy. If other types of germ cell tumors are present, either mixed or not mixed with germinoma, the outlook is usually not as good.

Other types of germ cell tumors (such as teratomas and yolk sac tumors) are rarely cured by surgery. Both radiation therapy and chemotherapy are used to treat them, but in some cases this might not control the tumor completely. Sometimes these tumors spread to the cerebrospinal fluid (CSF), and radiation therapy to the spinal cord and brain is needed as well.

Hyperlinks

- 1. <u>www.cancer.org/cancer/types/brain-spinal-cord-tumors-children/about/types-of-brain-and-spinal-tumors.html</u>
- 2. <u>www.cancer.org/cancer/types/brain-spinal-cord-tumors-children/causes-risks-prevention/risk-factors.html</u>
- 3. <u>www.cancer.org/cancer/types/brain-spinal-cord-tumors-children/detection-</u> <u>diagnosis-staging/how-diagnosed.html</u>
- 4. <u>www.cancer.org/cancer/types/brain-spinal-cord-tumors-children/detection-</u> <u>diagnosis-staging/how-diagnosed.html</u>
- 5. <u>www.cancer.org/cancer/managing-cancer/making-treatment-decisions/clinical-</u> <u>trials.html</u>
- 6. <u>www.cancer.org/cancer/managing-cancer/making-treatment-decisions/clinical-</u> <u>trials.html</u>
- 7. <u>www.cancer.org/cancer/types/brain-spinal-cord-tumors-children/causes-risks-prevention/risk-factors.html</u>
- 8. www.cancer.org/cancer/diagnosis-staging/tests/imaging-tests/mri-for-cancer.html
- 9. <u>www.cancer.org/cancer/types/brain-spinal-cord-tumors-children/causes-risks-prevention/risk-factors.html</u>
- 10. <u>www.cancer.org/cancer/managing-cancer/making-treatment-decisions/clinical-</u> <u>trials.html</u>
- 11. www.cancer.org/cancer/types/brain-spinal-cord-tumors-children/about/types-of-

brain-and-spinal-tumors.html

- 12. <u>www.cancer.org/cancer/types/brain-spinal-cord-tumors-children/detection-</u> <u>diagnosis-staging/staging.html</u>
- 13. <u>www.cancer.org/cancer/managing-cancer/treatment-types/stem-cell-</u> <u>transplant.html</u>
- 14. <u>www.cancer.org/cancer/managing-cancer/treatment-types/stem-cell-</u> <u>transplant.html</u>

References

Chang SM, Mehta MP, Vogelbaum MA, Taylor MD, Ahluwalia MS. Chapter 97: Neoplasms of the central nervous system. In: DeVita VT, Lawrence TS, Rosenberg SA, eds. *DeVita, Hellman, and Rosenberg's Cancer: Principles and Practice of Oncology*. 10th ed. Philadelphia, Pa: Lippincott Williams & Wilkins; 2015.

Dorsey JF, Hollander AB, Alonso-Basanta M, et al. Chapter 66: Cancer of the central nervous system. In: Abeloff MD, Armitage JO, Niederhuber JE. Kastan MB, McKenna WG, eds. *Abeloff's Clinical Oncology*. 5th ed. Philadelphia, Pa: Elsevier; 2014.

National Cancer Institute. Childhood Brain and Spinal Cord Tumors Treatment Overview (PDQ®). 2017. Accessed at www.cancer.gov/types/brain/hp/child-braintreatment-pdq#section/all on April 26, 2018.

Williams D, Parsons IF, Pollack DA. Chapter 26A: Gliomas, Ependymomas, and Other Nonembryonal Tumors of the Central Nervous System. In: Pizzo PA, Poplack DG, eds. *Principles and Practice of Pediatric Oncology*. 7th ed. Philadelphia, Pa: Lippincott Williams & Wilkins; 2016.

Last Revised: August 8, 2024

Written by

The American Cancer Society medical and editorial content team (<u>https://www.cancer.org/cancer/acs-medical-content-and-news-staff.html</u>)

Our team is made up of doctors and oncology certified nurses with deep knowledge of cancer care as well as editors and translators with extensive experience in medical

writing.

American Cancer Society medical information is copyrighted material. For reprint requests, please see our Content Usage Policy (www.cancer.org/aboutus/policies/content-usage.html).

cancer.org | 1.800.227.2345