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PDQ Cancer Information Summaries [Internet]. Bethesda (MD): National Cancer Institute (US); 2002-.

Childhood Ependymoma Treatment (PDQ®)

Patient Version

Authors

PDQ Pediatric Treatment Editorial Board.

Published online: April 6, 2020.

This PDQ cancer information summary has current information about the treatment of childhood ependymoma. It is meant to inform and help patients, families, and caregivers. It does not give formal guidelines or recommendations for making decisions about health care.

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General Information About Childhood Ependymoma

Key Points for This Section

- Childhood ependymoma is a disease in which malignant (cancer) cells form in the tissues of the brain and spinal cord.
- There are different types of ependymomas.
- The part of the brain that is affected depends on where the ependymoma forms.
- The cause of most childhood brain tumors is unknown.
- The signs and symptoms of childhood ependymoma are not the same in every child.
- Tests that examine the brain and spinal cord are used to detect (find) childhood ependymoma.
- Childhood ependymoma is diagnosed and removed in surgery.
- Certain factors affect prognosis (chance of recovery) and treatment options.

Childhood ependymoma is a disease in which malignant (cancer) cells form in the tissues of the brain and spinal cord.

The brain controls vital functions such as memory and learning, emotion, and the senses (hearing, sight, smell, taste, and touch). The spinal cord is made up of bundles of nerve fibers that connect the brain with nerves in most parts of the body.

Ependymomas form from ependymal cells that line the ventricles and passageways in the brain and the spinal cord. Ependymal cells make cerebrospinal fluid (CSF).

This summary is about the treatment of primary brain tumors (tumors that begin in the brain). Treatment of metastatic

brain tumors, which are tumors that begin in other parts of the body and spread to the brain, is not discussed in this summary.

There are many different types of brain tumors. Brain tumors can occur in both children and adults. However, treatment for children is different than treatment for adults. See the following PDQ summaries for more information:

- Childhood Brain and Spinal Cord Tumors Treatment Overview
- Adult Central Nervous System Tumors Treatment

There are different types of ependymomas.

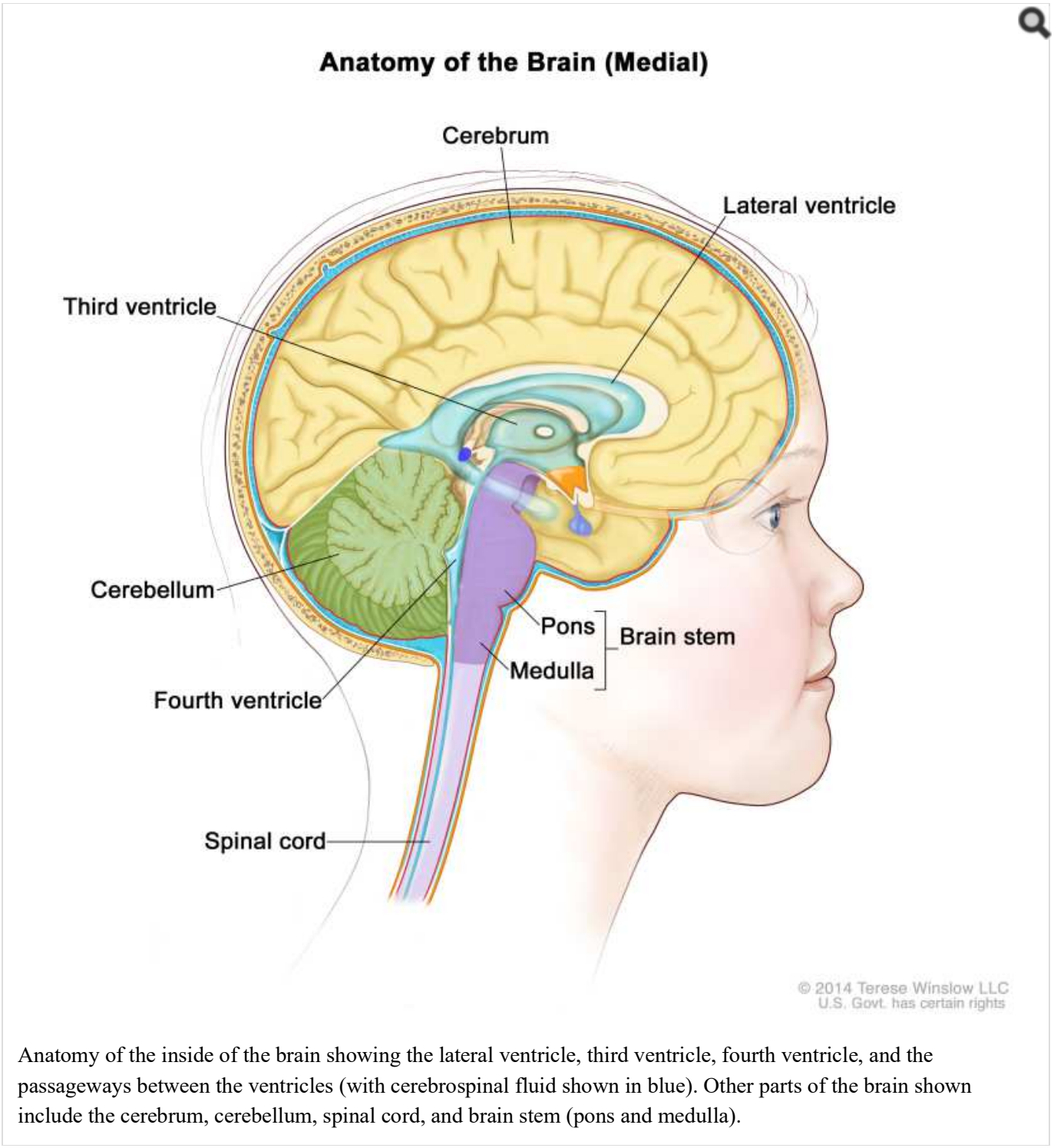
The World Health Organization (WHO) groups ependymal tumors into five main subtypes:

- Subependymoma (WHO grade I; rare in children).
- Myxopapillary ependymoma (WHO grade I).
- Ependymoma (WHO grade II).
- *RELA* fusion–positive ependymoma (WHO grade II or grade III with change in the *RELA* gene).
- Anaplastic ependymoma (WHO grade III).

The grade of a tumor describes how abnormal the cancer cells look under a microscope and how quickly the tumor is likely to grow and spread. Low-grade (grade I) cancer cells look more like normal cells than high-grade cancer cells (grade II and III). Grade I cancer cells also tend to grow and spread more slowly than grade II and III cancer cells.

The part of the brain that is affected depends on where the ependymoma forms.

Ependymomas can form anywhere in the fluid-filled ventricles and passageways in the brain and spinal cord. Most ependymomas form in the fourth ventricle and affect the cerebellum and the brain stem. Ependymomas form less commonly in the cerebrum and rarely in the spinal cord.



Anatomy of the inside of the brain showing the lateral ventricle, third ventricle, fourth ventricle, and the passageways between the ventricles (with cerebrospinal fluid shown in blue). Other parts of the brain shown include the cerebrum, cerebellum, spinal cord, and brain stem (pons and medulla).

Where the ependymoma forms affects the function of the brain and spinal cord:

- **Cerebellum**: The lower, back part of the brain (near the middle of the back of the head). The cerebellum controls movement, balance, and posture.
- **Brain stem**: The part that connects the brain to the spinal cord, in the lowest part of the brain (just above the back of the neck). The brain stem controls breathing, heart rate, and the nerves and muscles used in seeing, hearing, walking, talking, and eating.
- **Cerebrum**: The largest part of the brain, at the top of the head. The cerebrum controls thinking, learning, problem-solving, speech, emotions, reading, writing, and voluntary movement.

- **Spinal cord**: The column of nerve tissue that runs from the brain stem down the center of the back. It is covered by three thin layers of tissue called membranes. The spinal cord and membranes are surrounded by the vertebrae (back bones). Spinal cord nerves carry messages between the brain and the rest of the body, such as a message from the brain to cause muscles to move or a message from the skin to the brain to feel touch.

The cause of most childhood brain tumors is unknown.

The signs and symptoms of childhood ependymoma are not the same in every child.

Signs and symptoms depend on the following:

- The child's age.
- Where the tumor has formed.

Signs and symptoms may be caused by childhood ependymoma or by other conditions. Check with your child's doctor if your child has any of the following:

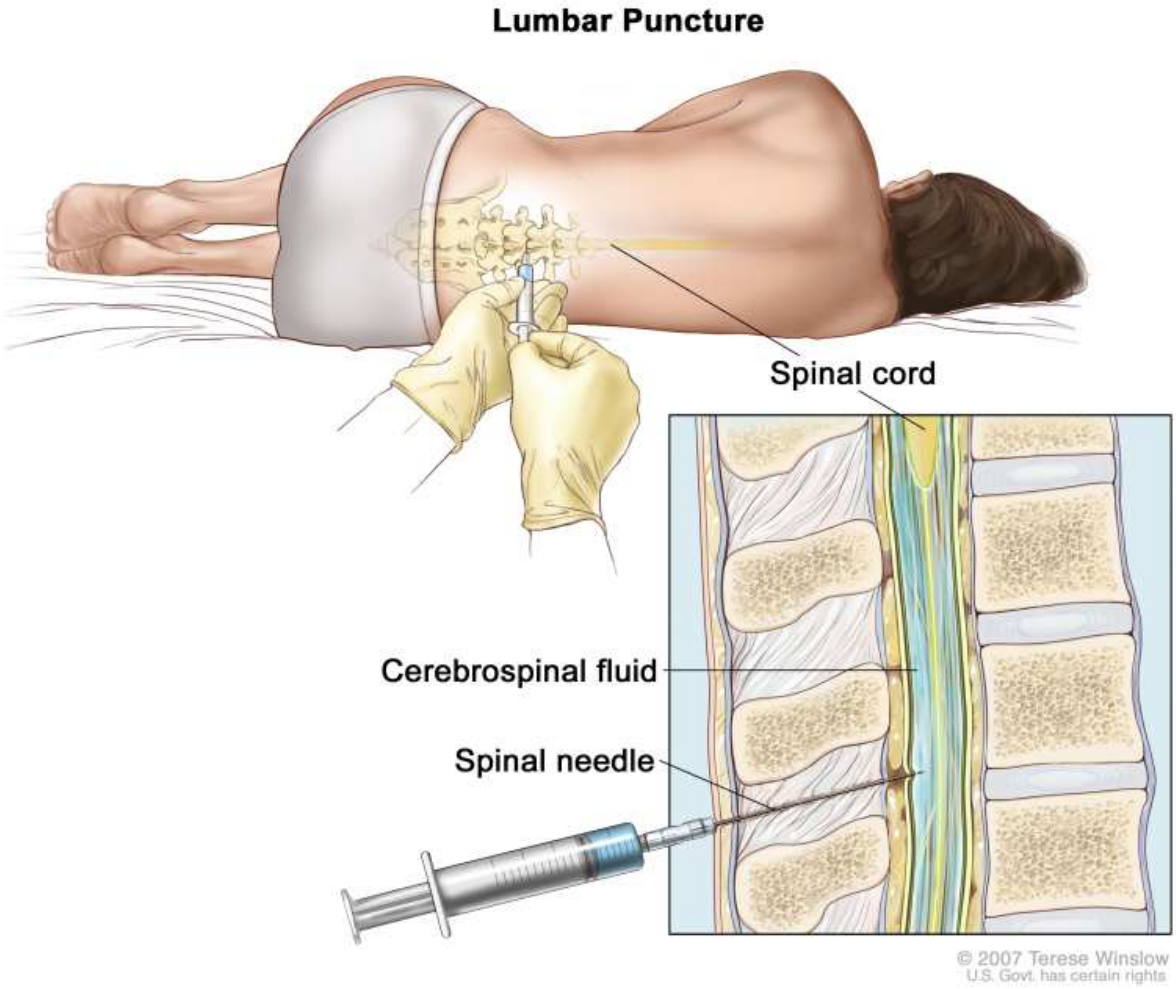
- Frequent headaches.
- Seizures.
- Nausea and vomiting.
- Pain in the neck or back.
- Loss of balance or trouble walking.
- Weakness in the legs.
- Blurry vision.
- A change in bowel function.
- Trouble urinating.
- Confusion or irritability.

Tests that examine the brain and spinal cord are used to detect (find) childhood ependymoma.

The following tests and procedures may be used:

- **Physical exam and health history**: An exam of the body to check general signs of health, including checking for signs of disease, such as lumps or anything else that seems unusual. A history of the patient's health habits and past illnesses and treatments will also be taken.
- **Neurological exam**: A series of questions and tests to check the brain, spinal cord, and nerve function. The exam checks a person's mental status, coordination, and ability to walk normally, and how well the muscles, senses, and reflexes work. This may also be called a neuro exam or a neurologic exam.
- **MRI (magnetic resonance imaging) with gadolinium**: A procedure that uses a magnet, radio waves, and a computer to make a series of detailed pictures of areas inside the brain and spinal cord. A substance called gadolinium is injected into a vein and travels through the bloodstream. The gadolinium collects around the cancer cells so they show up brighter in the picture. This procedure is also called nuclear magnetic resonance imaging (NMRI).
- **Lumbar puncture**: A procedure used to collect cerebrospinal fluid (CSF) from the spinal column. This is done

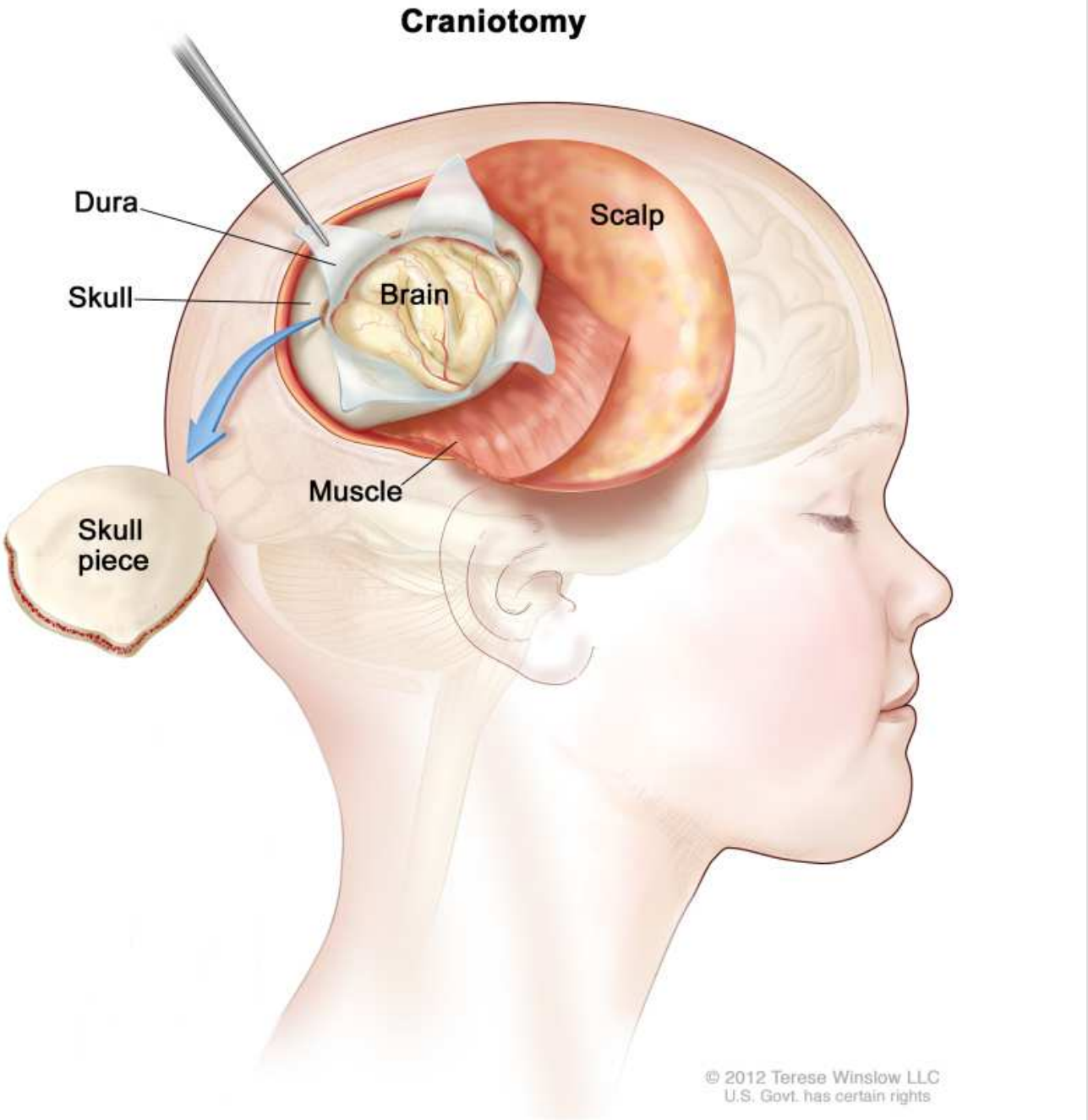
by placing a needle between two bones in the spine and into the CSF around the spinal cord and removing a sample of fluid. The sample of CSF is checked under a microscope for signs of tumor cells. The sample may also be checked for the amounts of protein and glucose. A higher than normal amount of protein or lower than normal amount of glucose may be a sign of a tumor. This procedure is also called an LP or spinal tap.



Lumbar puncture. A patient lies in a curled position on a table. After a small area on the lower back is numbed, a spinal needle (a long, thin needle) is inserted into the lower part of the spinal column to remove cerebrospinal fluid (CSF, shown in blue). The fluid may be sent to a laboratory for testing.

Childhood ependymoma is diagnosed and removed in surgery.

If the diagnostic tests show there may be a brain tumor, a biopsy is done by removing part of the skull and using a needle to remove a sample of the brain tissue. A pathologist views the tissue under a microscope to look for cancer cells and determine the grade of the tumor. If cancer cells are found, the doctor will remove as much tumor as safely possible during the same surgery.



Craniotomy: An opening is made in the skull and a piece of the skull is removed to show part of the brain.

The following test may be done on the tissue that was removed:

- **Immunohistochemistry**: A laboratory test that uses antibodies to check for certain antigens (markers) in a sample of a patient's tissue. The antibodies are usually linked to an enzyme or a fluorescent dye. After the antibodies bind to a specific antigen in the tissue sample, the enzyme or dye is activated, and the antigen can then be seen under a microscope. This type of test is used to help diagnose cancer and to help tell one type of cancer from another type of cancer.

Certain factors affect prognosis (chance of recovery) and treatment options.

The prognosis and treatment options depend on:

- Where the tumor has formed in the central nervous system (CNS).
- Whether there are certain changes in the genes or chromosomes.
- Whether any cancer cells remain after surgery to remove the tumor.
- The type and grade of ependymoma.
- The age of the child when the tumor is diagnosed.
- Whether the cancer has spread to other parts of the brain or spinal cord.
- Whether the tumor has just been diagnosed or has recurred (come back).

Prognosis also depends on whether radiation therapy was given, the type and treatment dose, and whether chemotherapy alone was given.

Stages of Childhood Ependymoma

Key Points for This Section

- There is no standard staging system for childhood ependymoma.
- Childhood ependymoma commonly comes back after it has been treated.

There is no standard staging system for childhood ependymoma.

Staging is the process used to find out whether cancer remains after surgery and if cancer has spread.

The treatment of ependymoma depends on the following:

- Where the cancer is in the brain or spinal cord.
- The age of the child.
- The type and grade of ependymoma.

Childhood ependymoma commonly comes back after it has been treated.

The tumor usually recurs (comes back) at the original cancer site. Childhood ependymoma may come back as long as 15 years or more after initial treatment.

Treatment Option Overview

Key Points for This Section

- There are different types of treatment for children with ependymoma.
- Children with ependymoma should have their treatment planned by a team of health care providers who are experts in treating childhood brain tumors.
- Three types of treatment are used:

- Surgery
- Radiation therapy
- Chemotherapy

- New types of treatment are being tested in clinical trials.
- Treatment for childhood ependymoma may cause side effects.
- Patients may want to think about taking part in a clinical trial.
- Patients can enter clinical trials before, during, or after starting their cancer treatment.
- Follow-up tests may be needed.

There are different types of treatment for children with ependymoma.

Different types of treatment are available for children with ependymoma. Some treatments are standard (the currently used treatment), and some are being tested in clinical trials. A treatment clinical trial is a research study meant to help improve current treatments or obtain information on new treatments for patients with cancer. When clinical trials show that a new treatment is better than the standard treatment, the new treatment may become the standard treatment.

Because cancer in children is rare, taking part in a clinical trial should be considered. Some clinical trials are open only to patients who have not started treatment.

Children with ependymoma should have their treatment planned by a team of health care providers who are experts in treating childhood brain tumors.

Treatment will be overseen by a pediatric oncologist, a doctor who specializes in treating children with cancer. The pediatric oncologist works with other pediatric health care providers who are experts in treating children with brain tumors and who specialize in certain areas of medicine. These may include the following specialists:

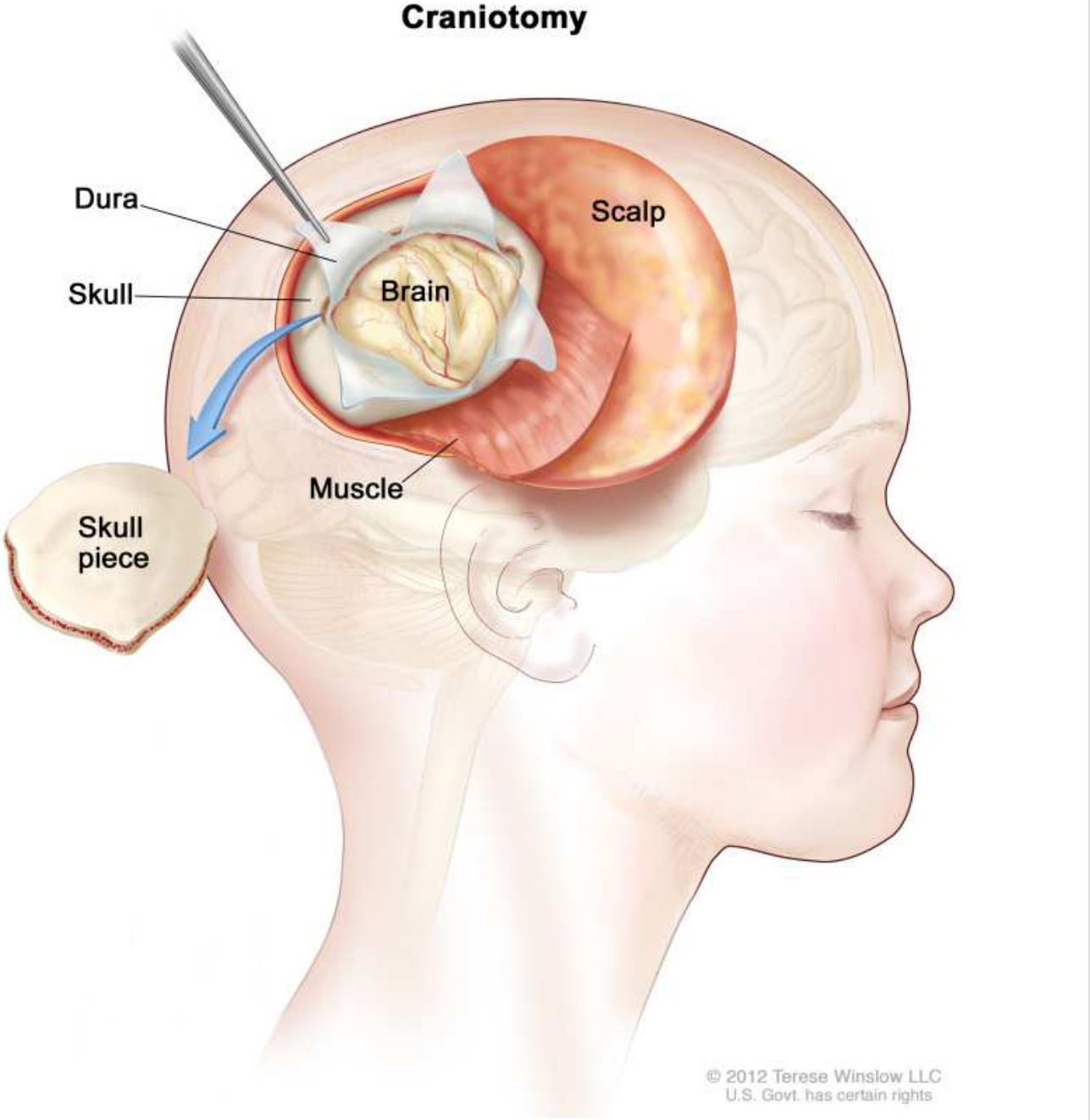
- Pediatric neurosurgeon.
- Neurologist.
- Pediatrician.
- Radiation oncologist.
- Medical oncologist.
- Endocrinologist.
- Rehabilitation specialist.
- Psychologist.
- Child-life specialist.

Three types of treatment are used:

Surgery

If the results of diagnostic tests show there may be a brain tumor, a biopsy is done by removing part of the skull and using a needle to remove a sample of the brain tissue. A pathologist views the tissue under a microscope to check for

cancer cells. If cancer cells are found, the doctor will remove as much tumor as safely possible during the same surgery.



Craniotomy: An opening is made in the skull and a piece of the skull is removed to show part of the brain.

An MRI is often done after the tumor is removed to find out whether any tumor remains. If tumor remains, a second surgery to remove as much of the remaining tumor as possible may be done.

After the doctor removes all the cancer that can be seen at the time of the surgery, some patients may be given chemotherapy or radiation therapy after surgery to kill any cancer cells that are left. Treatment given after surgery, to lower the risk that the cancer will come back, is called adjuvant therapy.

Radiation therapy

Radiation therapy is a cancer treatment that uses high-energy x-rays or other types of radiation to kill cancer cells or keep them from growing. External radiation therapy uses a machine outside the body to send radiation toward the area of the body with cancer.

Certain ways of giving radiation therapy can help keep radiation from damaging nearby healthy tissue. These types of radiation therapy include the following:

- Conformal radiation therapy: Conformal radiation therapy is a type of external radiation therapy that uses a computer to make a 3-dimensional (3-D) picture of the tumor and shapes the radiation beams to fit the tumor.
- Intensity-modulated radiation therapy (IMRT): IMRT is a type of 3-dimensional (3-D) radiation therapy that uses a computer to make pictures of the size and shape of the tumor. Thin beams of radiation of different intensities (strengths) are aimed at the tumor from many angles.
- Proton-beam radiation therapy: Proton-beam therapy is a type of high-energy, external radiation therapy. A radiation therapy machine aims streams of protons (tiny, invisible, positively-charged particles) at the cancer cells to kill them.
- Stereotactic radiosurgery: Stereotactic radiosurgery is a type of external radiation therapy. A rigid head frame is attached to the skull to keep the head still during the radiation treatment. A machine aims a single large dose of radiation directly at the tumor. This procedure does not involve surgery. It is also called stereotaxic radiosurgery, radiosurgery, and radiation surgery.

Younger children who receive radiation therapy to the brain have a higher risk of problems with growth and development than older children. 3-D conformal radiation therapy and proton-beam therapy are being studied in young children to see if the effects of radiation on growth and development are lessened.

Chemotherapy

Chemotherapy is a cancer treatment that uses drugs to stop the growth of cancer cells, either by killing the cells or by stopping them from dividing. When chemotherapy is taken by mouth or injected into a vein or muscle, the drugs enter the bloodstream and can reach cancer cells throughout the body (systemic chemotherapy).

New types of treatment are being tested in clinical trials.

This summary section describes treatments that are being studied in clinical trials. It may not mention every new treatment being studied. Information about clinical trials is available from the NCI website.

Targeted therapy

Targeted therapy is a type of treatment that uses drugs or other substances to attack cancer cells. Targeted therapies usually cause less harm to normal cells than chemotherapy or radiation therapy do.

Targeted therapy is being studied for the treatment of childhood ependymoma that has recurred (come back).

Treatment for childhood ependymoma may cause side effects.

For information about side effects that begin during treatment for cancer, see our Side Effects page.

Side effects from cancer treatment that begin after treatment and continue for months or years are called late effects. Late effects of cancer treatment may include the following:

- Physical problems, including problems with:

  - Tooth development.
  - Hearing function.
  - Bone and muscle growth and development.
  - Thyroid function.
  - Stroke.
- Changes in mood, feelings, thinking, learning, or memory.
- Second cancers (new types of cancer), such as thyroid cancer or brain cancer.

Some late effects may be treated or controlled. It is important to talk with your child's doctors about the effects cancer treatment can have on your child. (See the PDQ summary on Late Effects of Treatment for Childhood Cancer for more information.)

Patients may want to think about taking part in a clinical trial.

For some patients, taking part in a clinical trial may be the best treatment choice. Clinical trials are part of the cancer research process. Clinical trials are done to find out if new cancer treatments are safe and effective or better than the standard treatment.

Many of today's standard treatments for cancer are based on earlier clinical trials. Patients who take part in a clinical trial may receive the standard treatment or be among the first to receive a new treatment.

Patients who take part in clinical trials also help improve the way cancer will be treated in the future. Even when clinical trials do not lead to effective new treatments, they often answer important questions and help move research forward.

Patients can enter clinical trials before, during, or after starting their cancer treatment.

Some clinical trials only include patients who have not yet received treatment. Other trials test treatments for patients whose cancer has not gotten better. There are also clinical trials that test new ways to stop cancer from recurring (coming back) or reduce the side effects of cancer treatment.

Clinical trials are taking place in many parts of the country. Information about clinical trials supported by NCI can be found on NCI's clinical trials search webpage. Clinical trials supported by other organizations can be found on the ClinicalTrials.gov website.

Follow-up tests may be needed.

Some of the tests that were done to diagnose the cancer or to find out the stage of the cancer may be repeated. Some tests will be repeated in order to see how well the treatment is working. Decisions about whether to continue, change, or stop treatment may be based on the results of these tests.

Some of the tests will continue to be done from time to time after treatment has ended. The results of these tests can show if your child's condition has changed or if the cancer has recurred (come back). These tests are sometimes called follow-up tests or check-ups.

Follow-up tests for childhood ependymoma include an MRI (magnetic resonance imaging) of the brain and spinal cord at the following intervals:

- First 2 to 3 years after treatment: Every 3 to 4 months.
- Four to 5 years after treatment: Every 6 months.

- More than 5 years after treatment: Once a year.

Treatment of Childhood Myxopapillary Ependymoma

For information about the treatments listed below, see the Treatment Option Overview section.

Treatment of newly diagnosed childhood myxopapillary ependymoma (grade I) is:

- Surgery. Sometimes radiation therapy is given after surgery.

Treatment of Childhood Ependymoma, Anaplastic Ependymoma, and *RELA* Fusion–positive Ependymoma

For information about the treatments listed below, see the Treatment Option Overview section.

Treatment of newly diagnosed childhood ependymoma (grade II), anaplastic ependymoma (grade III), and *RELA* fusion–positive ependymoma (grade II or grade III) is:

- Surgery.

After surgery, the plan for further treatment depends on the following:

- Whether any cancer cells remain after surgery.
- Whether the cancer has spread to other parts of the brain or spinal cord.
- The age of the child.

When the tumor is completely removed and cancer cells have not spread, treatment may include the following:

- Radiation therapy.

When part of the tumor remains after surgery, but cancer cells have not spread, treatment may include the following:

- A second surgery to remove as much of the remaining tumor as possible.
- Radiation therapy.
- Chemotherapy.

When cancer cells have spread within the brain and spinal cord, treatment may include the following:

- Radiation therapy to the brain and spinal cord.
- Chemotherapy.

Treatment for children younger than 1 year of age may include the following:

- Chemotherapy.
- Radiation therapy. Radiation therapy is not given to children until they are older than 1 year of age.
- A clinical trial of 3-dimensional (3-D) conformal radiation therapy or proton-beam radiation therapy.

Treatment of Recurrent Childhood Ependymoma

For information about the treatments listed below, see the Treatment Option Overview section.

Treatment of recurrent childhood ependymoma may include the following:

- Surgery.
- Radiation therapy, which may include stereotactic radiosurgery, intensity-modulated radiation therapy, or proton-beam radiation therapy.
- Chemotherapy.
- A clinical trial that checks a sample of the patient's tumor for certain gene changes. The type of targeted therapy that will be given to the patient depends on the type of gene change.
- A clinical trial of surgery and repeat radiation therapy to treat ependymoma that has come back after initial treatment.

To Learn More About Childhood Brain Tumors

For more information about childhood brain tumors, see the following:

- Pediatric Brain Tumor Consortium (PBTC)
- Targeted Cancer Therapies

For more childhood cancer information and other general cancer resources, see the following:

- About Cancer
- Childhood Cancers
- CureSearch for Children's Cancer
- Late Effects of Treatment for Childhood Cancer
- Adolescents and Young Adults with Cancer
- Children with Cancer: A Guide for Parents
- Cancer in Children and Adolescents
- Staging
- Coping with Cancer
- Questions to Ask Your Doctor about Cancer
- For Survivors and Caregivers

About This PDQ Summary

About PDQ

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Clinical Trial Information

A clinical trial is a study to answer a scientific question, such as whether one treatment is better than another. Trials are based on past studies and what has been learned in the laboratory. Each trial answers certain scientific questions in order to find new and better ways to help cancer patients. During treatment clinical trials, information is collected about the effects of a new treatment and how well it works. If a clinical trial shows that a new treatment is better than one currently being used, the new treatment may become "standard." Patients may want to think about taking part in a clinical trial. Some clinical trials are open only to patients who have not started treatment.

Clinical trials can be found online at NCI's website. For more information, call the Cancer Information Service (CIS), NCI's contact center, at 1-800-4-CANCER (1-800-422-6237).

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