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Childhood Brain Stem Glioma Treatment (PDQ®)

Patient Version

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This PDQ cancer information summary has current information about the treatment of childhood brain stem glioma. 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 Brain Stem Glioma

Key Points for This Section

- Childhood brain stem glioma is a disease in which benign (noncancer) or malignant (cancer) cells form in the tissues of the brain stem.
- There are two types of brain stem gliomas in children.
- The cause of most childhood brain tumors is unknown.
- The signs and symptoms of brain stem glioma are not the same in every child.
- Tests that examine the brain are used to detect (find) childhood brain stem glioma.
- A biopsy may be done to diagnose certain types of brain stem glioma.
- Certain factors affect prognosis (chance of recovery).

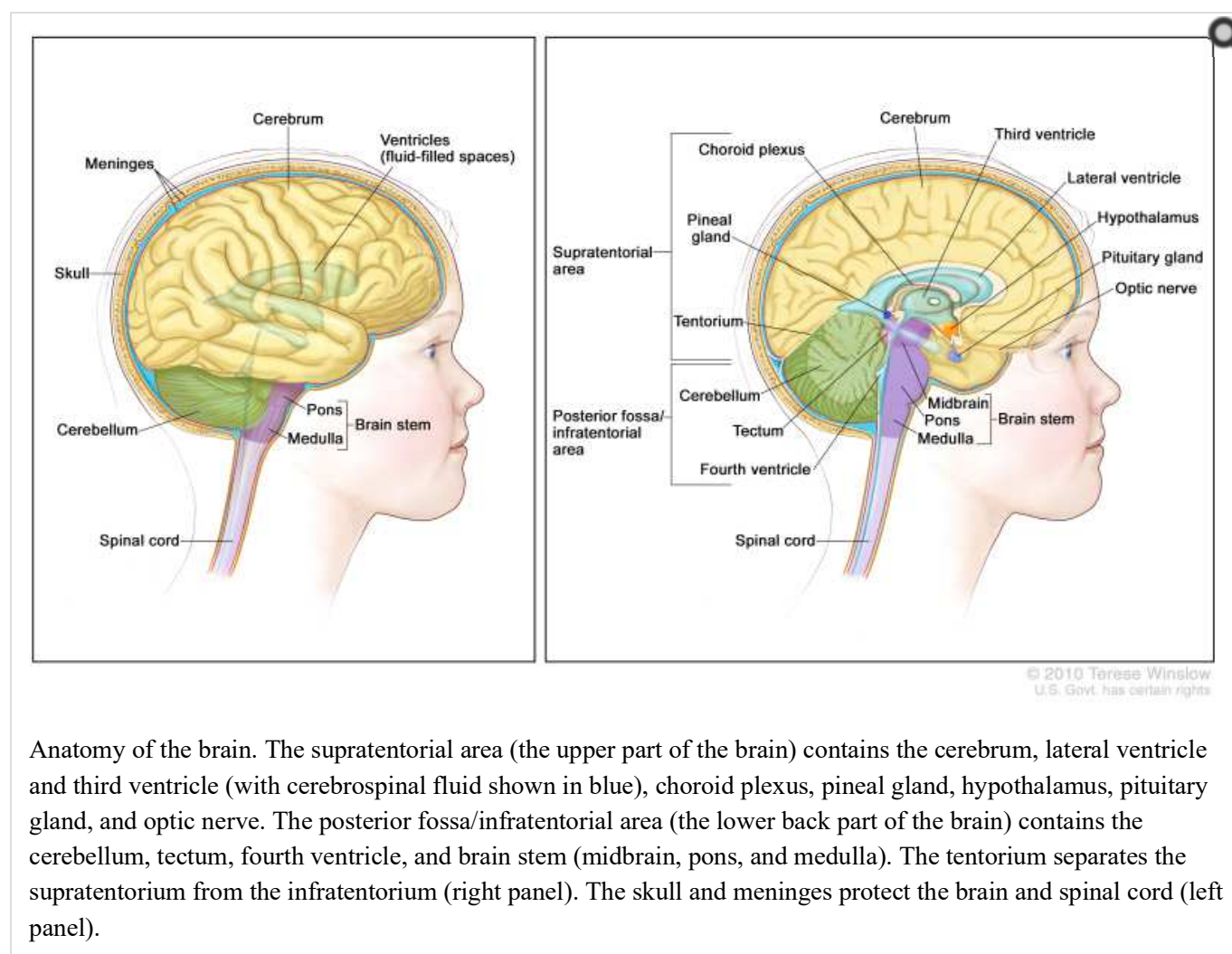
Childhood brain stem glioma is a disease in which benign (noncancer) or malignant (cancer) cells form in the tissues of the brain stem.

Gliomas are tumors formed from glial cells. Glial cells in the brain hold nerve cells in place, bring food and oxygen to nerve cells, and help protect nerve cells from disease, such as infection. In brain stem glioma, the glial cells in the brain stem are affected.

The brain stem is made up of the midbrain, pons, and medulla. It is the lowest part of the brain and connects to the spinal cord, just above the back of the neck. The brain stem controls vital functions such as breathing, heart rate, and blood pressure, and the nerves and muscles used in seeing, hearing, walking, talking, and eating.

Most childhood brain stem gliomas are diffuse intrinsic pontine gliomas (DIPG), which form in the pons. Focal

gliomas form in other parts of the brain stem.



Anatomy of the brain. The supratentorial area (the upper part of the brain) contains the cerebrum, lateral ventricle and third ventricle (with cerebrospinal fluid shown in blue), choroid plexus, pineal gland, hypothalamus, pituitary gland, and optic nerve. The posterior fossa/infratentorial area (the lower back part of the brain) contains the cerebellum, tectum, fourth ventricle, and brain stem (midbrain, pons, and medulla). The tentorium separates the supratentorium from the infratentorium (right panel). The skull and meninges protect the brain and spinal cord (left panel).

Brain tumors are the second most common type of cancer in children.

This summary is about the treatment of primary brain tumors (tumors that begin in the brain). Treatment for metastatic brain tumors, which are tumors formed by cancer cells that begin in other parts of the body and spread to the brain, is not discussed in this summary.

Brain tumors can occur in both children and adults; however, treatment for children may be different than treatment for adults. For information on treatment of brain tumors in adults, see the PDQ summary [Adult Central Nervous System Tumors Treatment](#).

There are two types of brain stem gliomas in children.

Even though DIPG and focal brain stem glioma form in the same type of cell, they act differently:

- **DIPG.** DIPG is a fast-growing tumor that forms in the pons. DIPG is hard to treat and has a poor prognosis (chance of recovery) because of the following:
 - It is not a well-defined tumor and spreads among the healthy cells in the brain stem.
 - Vital functions, such as breathing and heart rate, may be affected.
- **Focal brain stem glioma.** A focal glioma is a slow-growing tumor that forms outside the pons and in only one

area of the brain stem. It is easier to treat and has a better prognosis than DIPG.

The cause of most childhood brain tumors is unknown.

Anything that increases your risk of getting a disease is called a risk factor. Having a risk factor does not mean that you will get cancer; not having risk factors doesn't mean that you will not get cancer. Talk with your child's doctor if you think your child may be at risk. Possible risk factors for brain stem glioma include:

- Having certain genetic disorders, such as neurofibromatosis type 1 (NF1).

The signs and symptoms of brain stem glioma are not the same in every child.

Signs and symptoms depend on the following:

- Where the tumor forms in the brain.
- The size of the tumor and whether it has spread throughout the brain stem.
- How fast the tumor grows.
- The child's age and stage of development.

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

- Trouble with eye movement (the eye is turned inward).
- Vision problems.
- Morning headache or headache that goes away after vomiting.
- Nausea and vomiting.
- Unusual sleepiness.
- Loss of ability to move one side of the face or body.
- Loss of balance and trouble walking.
- More or less energy than usual.
- Changes in behavior.
- Trouble learning in school.

Tests that examine the brain are used to detect (find) childhood brain stem glioma.

The following tests and procedures may be used:

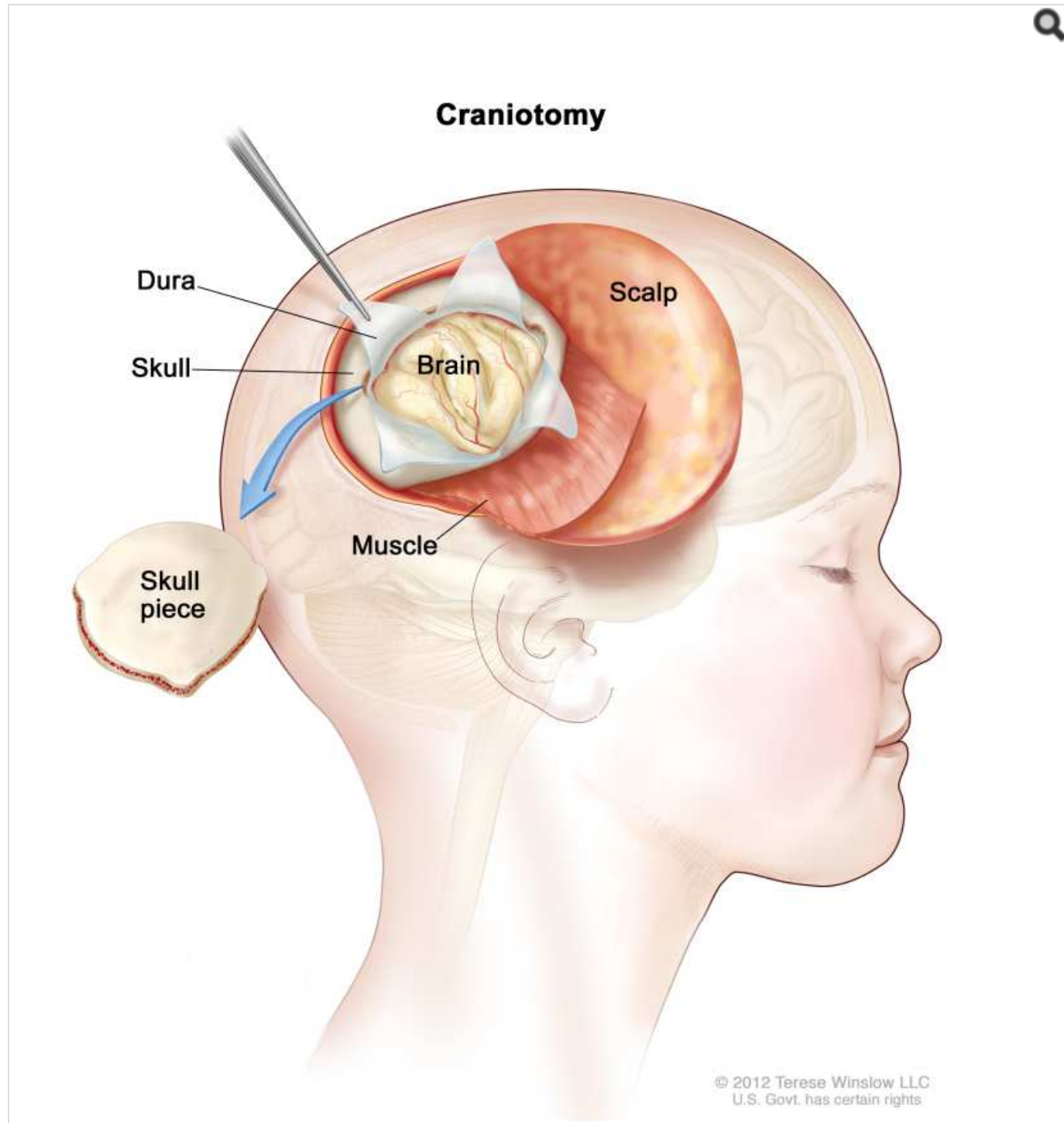
- **Physical exam and health history:** An exam of the body to check general signs of health, including checking for signs of disease, such as lumps or anything else that seems unusual. A history of the patient's health habits and past illnesses and treatments will also be taken.
- **Neurological exam:** 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. A substance called gadolinium is

injected into a vein. 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).

A biopsy may be done to diagnose certain types of brain stem glioma.

If the MRI scan looks like the tumor is a DIPG, a biopsy is usually not done and the tumor is not removed. When the MRI scan results are uncertain, a biopsy may be done.

If the MRI scan looks like a focal brain stem glioma, a biopsy may be done. A part of the skull is removed and a needle is used to remove a sample of the brain tissue. Sometimes, the needle is guided by a computer. A pathologist views the tissue under a microscope to look 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.

The following test may be done on the tissue sample that was removed during biopsy or surgery:

- **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).

The child's prognosis depends on the following:

- The type of brain stem glioma (DIPG or focal glioma).
- Where the tumor is found in the brain and if it has spread within the brain stem.
- The age of the child at diagnosis.
- How long the child has symptoms prior to diagnosis.
- Whether or not the child has a condition called neurofibromatosis type 1.
- Whether there is a certain change in the H3 K27m gene.
- Whether the tumor has just been diagnosed or has recurred (come back).

Most children with DIPG live less than 18 months after diagnosis. Children with a focal glioma usually live longer than 5 years.

Stages of Childhood Brain Stem Glioma

Key Points for This Section

- The plan for cancer treatment depends on whether the tumor is in one area of the brain or has spread all through the brain.

The plan for cancer treatment depends on whether the tumor is in one area of the brain or has spread all through the brain.

Staging is the process used to find out how much cancer there is and if cancer has spread. It is important to know the stage in order to plan treatment.

There is no standard staging system for childhood brain stem glioma. Treatment is based on the following:

- Whether the tumor is newly diagnosed or recurrent (has come back after treatment).
- The type of tumor (either a diffuse intrinsic pontine glioma or a focal glioma).

A focal brain stem glioma may recur many years after first being treated. The tumor may come back in the brain or in other parts of the central nervous system. Before cancer treatment is given, imaging tests, a biopsy, or surgery may be

done to make sure there is cancer and find out how much cancer there is.

Treatment Option Overview

Key Points for This Section

- There are different types of treatment for children with brain stem glioma.
- Children with brain stem glioma should have their treatment planned by a team of health care providers who are experts in treating childhood brain tumors.
- Five types of standard treatment are used:
 - Surgery
 - Radiation therapy
 - Chemotherapy
 - Cerebrospinal fluid diversion
 - Observation
- New types of treatment are being tested in clinical trials.
- Treatment for childhood brain stem glioma 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 brain stem glioma.

Different types of treatment are available for children with brain stem glioma. 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 brain stem glioma 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:

- Pediatrician.
- Neurosurgeon.

- [Neuropathologist](#).
- [Pediatric radiation oncologist](#).
- [Neuro-oncologist](#).
- [Neurologist](#).
- [Neuroradiologist](#).
- [Endocrinologist](#).
- [Psychologist](#).
- [Rehabilitation specialist](#).
- [Social worker](#).
- [Child-life specialist](#).

Five types of standard treatment are used:

Surgery

[Biopsy](#) or [surgery](#) to remove DIPG is not usually done because of the following:

- DIPG is not a single [mass](#). It spreads between the healthy brain [cells](#) in the [brain stem](#).
- [Vital functions](#), such as breathing and [heart rate](#) may be affected.

A biopsy to [diagnose](#) or surgery to remove the tumor may be used for childhood [focal brain stem glioma](#).

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. There are two types of radiation therapy:

- [External radiation therapy](#) uses a machine outside the body to send radiation toward the area of the body with cancer.
- [Internal radiation therapy](#) uses a [radioactive substance](#) sealed in [needles](#), [seeds](#), [wires](#), or [catheters](#) that are placed directly into or near the cancer.

The way the radiation therapy is given depends on the type of the cancer being treated. External radiation therapy is used to treat DIPG. External and/or internal radiation therapy may be used to treat [focal brain stem gliomas](#).

Several months after radiation therapy to the brain, [imaging tests](#) may show changes to the brain [tissue](#). These changes may be caused by the radiation therapy or may mean the tumor is growing. It is important to be sure the tumor is growing before any more treatment is given.

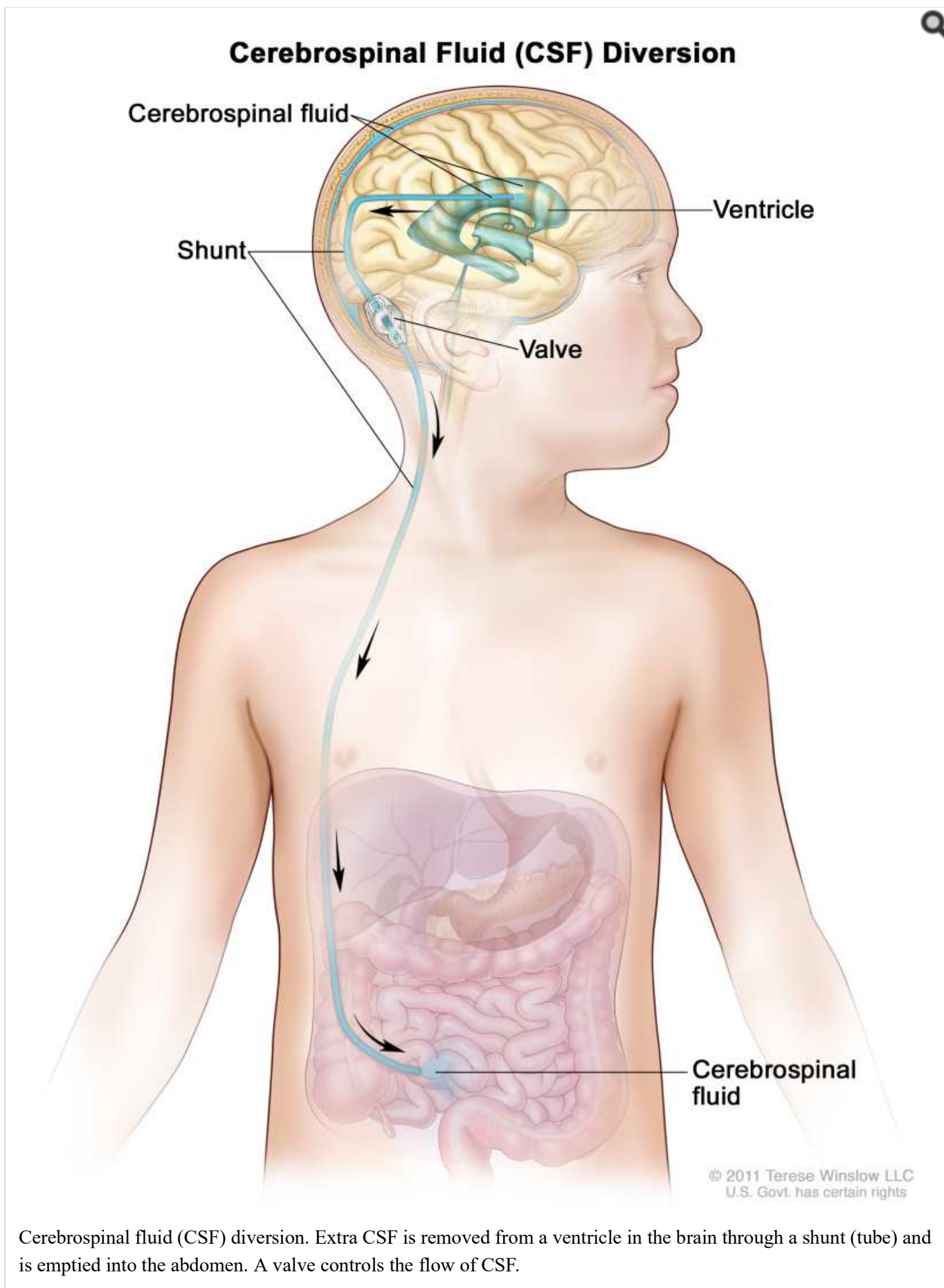
Chemotherapy

[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](#)).

Because radiation therapy to the brain can affect growth and brain development in young children, chemotherapy may be given to delay or reduce the need for radiation therapy.

Cerebrospinal fluid diversion

Cerebrospinal fluid diversion is a method used to drain fluid that has built up in the brain. A shunt (long, thin tube) is placed in a ventricle (fluid-filled space) of the brain and threaded under the skin to another part of the body, usually the abdomen. The shunt carries extra fluid away from the brain so it may be absorbed elsewhere in the body.



Observation

Observation is closely monitoring a patient's condition without giving any treatment until signs or symptoms appear or change.

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 may cause less harm to normal cells than chemotherapy or radiation therapy do.

There are different types of targeted therapy being studied in the treatment of brain stem gliomas:

- Kinase inhibitor therapy blocks certain proteins, such as BRAF or MEK, which may help keep cancer cells from growing or dividing. Dabrafenib (BRAF kinase inhibitor) and trametinib (MEK kinase inhibitor) are being studied to treat newly diagnosed focal glioma and recurrent brain stem glioma.
- Histone deacetylase inhibitor (HDI) therapy may stop the growth of tumor cells by blocking some of the enzymes needed for cell growth. It is also a type of angiogenesis agent. Panobinostat is being studied in the treatment of DIPG that did not respond to treatment or recurred.
- Monoclonal antibody therapy uses antibodies made in the laboratory from a single type of immune system cell. These antibodies can identify substances on cancer cells or normal substances that may help cancer cells grow. The antibodies attach to the substances and kill the cancer cells, block their growth, or keep them from spreading.

A monoclonal antibody, APX005M, binds to CD40, a cell surface receptor found on certain immune cells and some cancer cells. It may fight cancer by boosting the immune system and by slowing cancer cell growth. It is being studied in the treatment of pediatric brain tumors that are growing, spreading, or getting worse (progressive), or in newly diagnosed DIPG.

Treatment for childhood brain stem glioma 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 may include the following:

- Physical problems.
- Changes in mood, feelings, thinking, learning, or memory.
- Second cancers (new types of 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.

Information about clinical trials is available from the [NCI website](#).

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.

If the results of imaging tests done after treatment for DIPG show a mass in the brain, a biopsy may be done to find out if it is made up of dead tumor cells or if new cancer cells are growing. In children who are expected to live a long time, regular MRIs may be done to see if the cancer has come back.

Treatment of DIPG

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

Newly diagnosed childhood diffuse intrinsic brain stem glioma (DIPG) is a tumor for which no treatment has been given. The child may have received drugs or treatment to relieve signs or symptoms caused by the tumor.

Standard treatment of DIPG may include the following:

- External radiation therapy.
- Chemotherapy (in infants).
- A clinical trial of a new treatment.

Treatment of Focal Brain Stem Glioma

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

Newly diagnosed childhood focal glioma is a tumor for which no treatment has been given. The child may have received drugs or treatment to relieve signs or symptoms caused by the tumor.

Treatment of [focal glioma](#) may include the following:

- [Surgery](#) to remove the tumor may be followed by [chemotherapy](#) and/or [external radiation therapy](#).
- [Observation](#) for small [tumors](#) that grow slowly. [Cerebrospinal fluid diversion](#) may be done when there is extra fluid in the brain.
- [Internal radiation therapy](#) with [radioactive seeds](#), with or without chemotherapy, when the tumor cannot be removed by surgery.
- A [clinical trial](#) of targeted therapy with a [BRAF kinase inhibitor](#) (dabrafenib) combined with a [MEK inhibitor](#) (trametinib), for certain tumors that cannot be removed by surgery.

Treatment of brain stem glioma in children with [neurofibromatosis type 1](#) may be [observation](#). The tumors are slow-growing in these children and may not need specific treatment for years.

Treatment of Progressive or Recurrent Childhood Brain Stem Glioma

When cancer does not get better with treatment or comes back, [palliative care](#) is an important part of the child's treatment plan. It includes [physical](#), [psychological](#), [social](#), and [spiritual](#) support for the child and family. The goal of palliative care is to help control symptoms and give the child the best quality of life possible. Parents may not be sure about whether to continue treatment or what kind of treatment is best for their child. The [healthcare team](#) can give parents information to help them make these decisions.

For information about the treatments listed below, see the [Treatment Option Overview](#) section.

More [radiation therapy](#) may be given to children with [progressive](#) or [recurrent diffuse intrinsic pontine glioma](#) (DIPG) who responded when first treated with radiation therapy. Treatment of progressive or recurrent DIPG may also include the following:

- 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 targeted therapy with a [histone deacetylase inhibitor](#) (panobinostat) or a [monoclonal antibody](#) (APX005M).

Treatment of recurrent [focal childhood brain stem glioma](#) may include the following:

- A second surgery to remove the tumor.
- [External 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.

To Learn More About Childhood Brain Tumors

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

- [Targeted Cancer Therapies](#)
- [Pediatric Brain Tumor Consortium \(PBTC\)](#)
- [Communication in Cancer Care](#)

- [Planning the Transition to End-of-Life Care in Advanced Cancer](#)
- [Pediatric Supportive Care \(End of Life Care\)](#)

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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Reviewers and Updates

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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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