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Childhood Brain and Spinal Cord Tumors Treatment Overview (PDQ®)

Patient Version

PDQ Pediatric Treatment Editorial Board.

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This PDQ cancer information summary has current information about the treatment of childhood brain and spinal cord tumors. 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.

Editorial Boards write the PDQ cancer information summaries and keep them up to date. These Boards are made up of experts in cancer treatment and other specialties related to cancer. The summaries are reviewed regularly and changes are made when there is new information. The date on each summary ("Date Last Modified") is the date of the most recent change. The information in this patient summary was taken from the health professional version, which is reviewed regularly and updated as needed, by the PDQ Pediatric Treatment Editorial Board.

General Information About Childhood Brain and Spinal Cord Tumors

Key Points for This Section

- A childhood brain or spinal cord tumor is a disease in which abnormal cells form in the tissues of the brain or spinal cord.
- The brain controls many important body functions.
- The spinal cord connects the brain with nerves in most parts of the body.
- Brain and spinal cord tumors are a common type of childhood cancer.
- The cause of most childhood brain and spinal cord tumors is unknown.
- The signs and symptoms of childhood brain and spinal cord tumors are not the same in every child.
- Tests that examine the brain and spinal cord are used to detect (find) childhood brain and spinal cord tumors.
- Most childhood brain tumors are diagnosed and removed in surgery.
- Some childhood brain and spinal cord tumors are diagnosed by imaging tests.
- Certain factors affect prognosis (chance of recovery).

A childhood brain or spinal cord tumor is a disease in which abnormal cells form in the tissues of the brain or spinal cord.

There are many types of childhood brain and spinal cord tumors. The tumors are formed by the abnormal growth of cells and may begin in different areas of the brain or spinal cord.

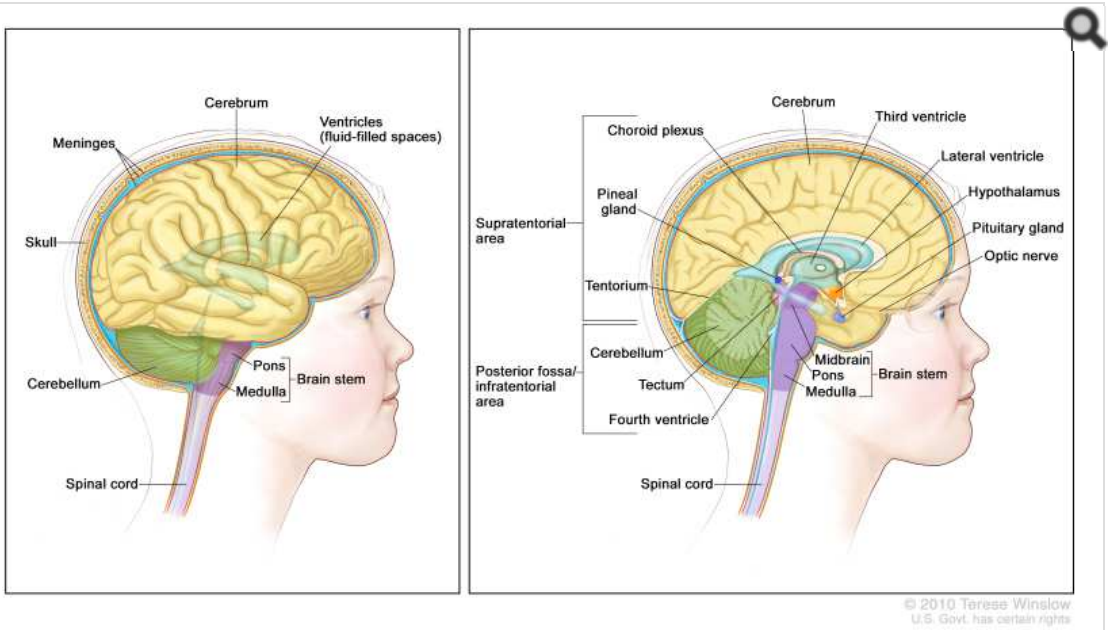
The tumors may be benign (not cancer) or malignant (cancer). Benign brain tumors grow and press on nearby areas of the brain. They rarely spread into other tissues. Malignant brain tumors are likely to grow quickly and spread into other brain tissue. When a tumor grows into or presses on an area of the brain, it may stop that part of the brain from working the way it should. Both benign and malignant brain tumors can cause signs or symptoms and need treatment.

Together, the brain and spinal cord make up the central nervous system (CNS).

The brain controls many important body functions.

The brain has three major parts:

- The cerebrum is the largest part of the brain. It is at the top of the head. The cerebrum controls thinking, learning, problem solving, emotions, speech, reading, writing, and voluntary movement.
- The cerebellum is in the lower back of the brain (near the middle of the back of the head). It controls movement, balance, and posture.
- The brain stem connects the brain to the spinal cord. It is 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.



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).

The spinal cord connects the brain with nerves in most parts of the body.

The spinal cord is a 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. These 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.

Brain and spinal cord tumors are a common type of childhood cancer.

Although cancer is rare in children, brain and spinal cord tumors are the second most common type of childhood cancer, after leukemia. Brain tumors can occur in both children and adults. Treatment for children is usually different than treatment for adults. (See the PDQ summary on Adult Central Nervous System Tumors Treatment for more information about the treatment of adults.)

This summary describes the treatment of primary brain and spinal cord tumors (tumors that begin in the brain and spinal cord). Treatment of metastatic brain and spinal cord tumors is not covered in this summary. Metastatic tumors are formed by cancer cells that begin in other parts of the body and spread to the brain or spinal cord.

The cause of most childhood brain and spinal cord tumors is unknown.

The signs and symptoms of childhood brain and spinal cord tumors are not the same in every child.

Signs and symptoms depend on the following:

- Where the tumor forms in the brain or spinal cord.
- The size of the tumor.
- How fast the tumor grows.
- The child's age and development.

Signs and symptoms may be caused by childhood brain and spinal cord tumors or by other conditions, including cancer that has spread to the brain. Check with your child's doctor if your child has any of the following:

Brain Tumor Signs and Symptoms

- Morning headache or headache that goes away after vomiting.
- Frequent nausea and vomiting.
- Vision, hearing, and speech problems.
- Loss of balance and trouble walking.
- Unusual sleepiness or change in activity level.
- Unusual changes in personality or behavior.
- Seizures.
- Increase in the head size (in infants).

Spinal Cord Tumor Signs and Symptoms

- Back pain or pain that spreads from the back towards the arms or legs.
- A change in bowel habits or trouble urinating.
- Weakness in the legs.
- Trouble walking.

In addition to these signs and symptoms of brain and spinal cord tumors, some children are unable to reach certain growth and development milestones such as sitting up, walking, and talking in sentences.

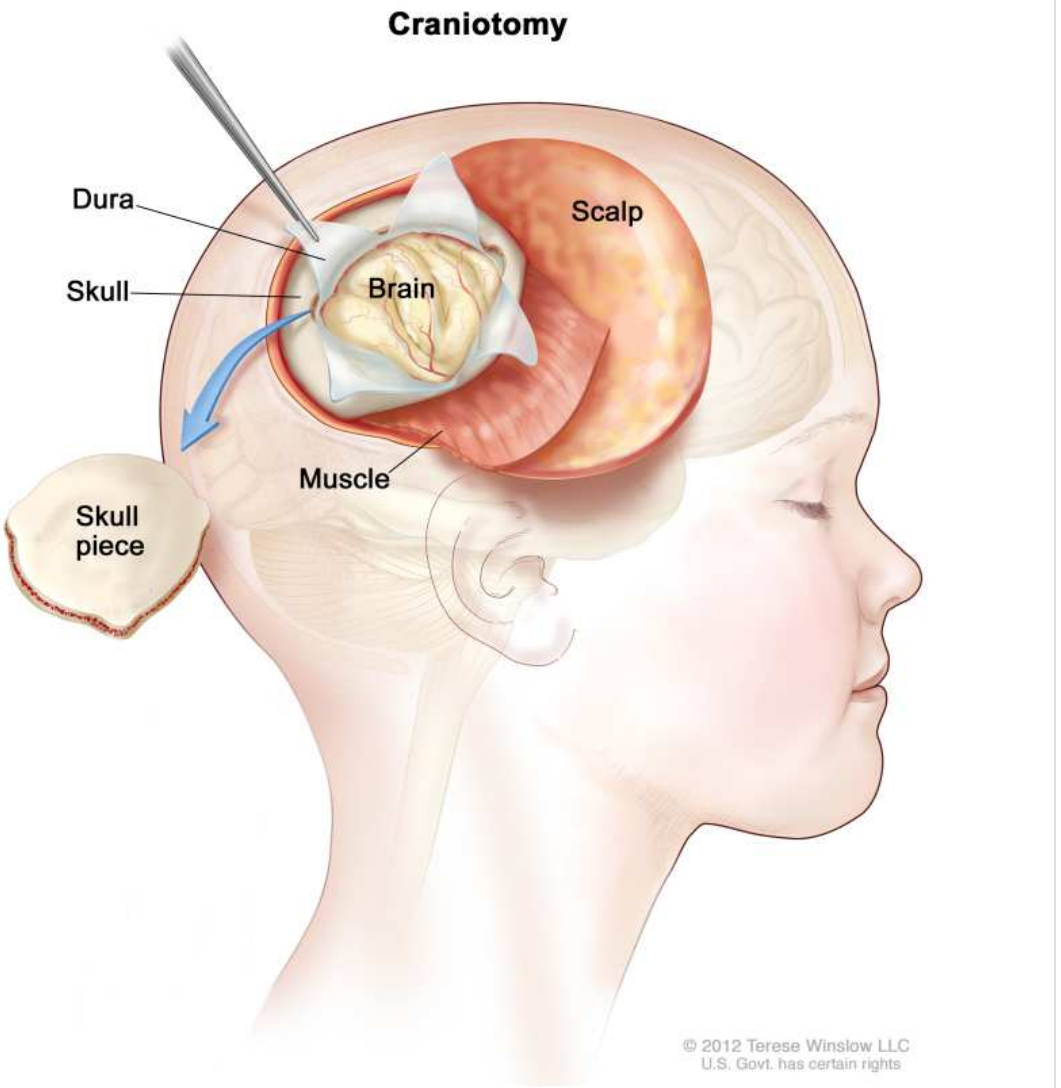
Tests that examine the brain and spinal cord are used to detect (find) childhood brain and spinal cord tumors.

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 the brain and spinal cord. 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).
- **Serum tumor marker test**: A procedure in which a sample of blood is examined to measure the amounts of certain substances released into the blood by organs, tissues, or tumor cells in the body. Certain substances are linked to specific types of cancer when found in increased levels in the blood. These are called tumor markers.

Most childhood brain tumors are diagnosed and removed in surgery.

If doctors think there might be a brain tumor, a biopsy may be done to remove a sample of tissue. For tumors in the brain, the biopsy is done by removing part of the skull and using a needle to remove a sample of tissue. A pathologist views the tissue under a microscope to look for cancer cells. If cancer cells are found, the doctor may remove as much tumor as safely possible during the same surgery. The pathologist checks the cancer cells to find out the type and grade of brain tumor. The grade of the tumor is based on how abnormal the cancer cells look under a microscope and how quickly the tumor is likely to grow and spread.



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 sample of tissue that is 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.

Some childhood brain and spinal cord tumors are diagnosed by imaging tests.

Sometimes a biopsy or surgery cannot be done safely because of where the tumor formed in the brain or spinal cord. These tumors are diagnosed based on the results of imaging tests and other procedures.

Certain factors affect prognosis (chance of recovery).

The prognosis depends on the following:

- Whether there are any cancer cells left after surgery.
- The type of tumor.
- Where the tumor is in the body.
- The child's age.
- Whether the tumor has just been diagnosed or has recurred (come back).

Staging Childhood Brain and Spinal Cord Tumors

Key Points for This Section

- In childhood brain and spinal cord tumors, treatment options are based on several factors.
- The information from tests and procedures done to detect (find) childhood brain and spinal cord tumors is used to determine the tumor risk group.
- Sometimes childhood brain and spinal cord tumors come back after treatment.

In childhood brain and spinal cord tumors, treatment options are based on several factors.

Staging is the process used to find how much cancer there is and if cancer has spread within the brain, spinal cord, or to other parts of the body. It is important to know the stage in order to plan cancer treatment.

In childhood brain and spinal cord tumors, there is no standard staging system. Instead, the plan for cancer treatment depends on several factors:

- The type of tumor and where the tumor formed in the brain.
- Whether the tumor is newly diagnosed or recurrent. A newly diagnosed brain or spinal cord tumor is one that has never been treated. A recurrent childhood brain or spinal cord tumor is one that has recurred (come back) after it has been treated. Childhood brain and spinal cord tumors may come back in the same place or in another part of the brain, or spinal cord. Sometimes they come back in another part of the body. The tumor may come back many years after first being treated. Tests and procedures, including biopsy, that were done to diagnose and stage the tumor may be done to find out if the tumor has recurred.
- The grade of the tumor. The grade of the tumor is based on how abnormal the cancer cells look under a microscope and how quickly the tumor is likely to grow and spread. It is important to know the grade of the tumor and if there were any cancer cells remaining after surgery in order to plan treatment. The grade of the tumor is not used to plan treatment for all types of brain and spinal cord tumors.
- The tumor risk group. Risk groups are either average risk and poor risk or low,

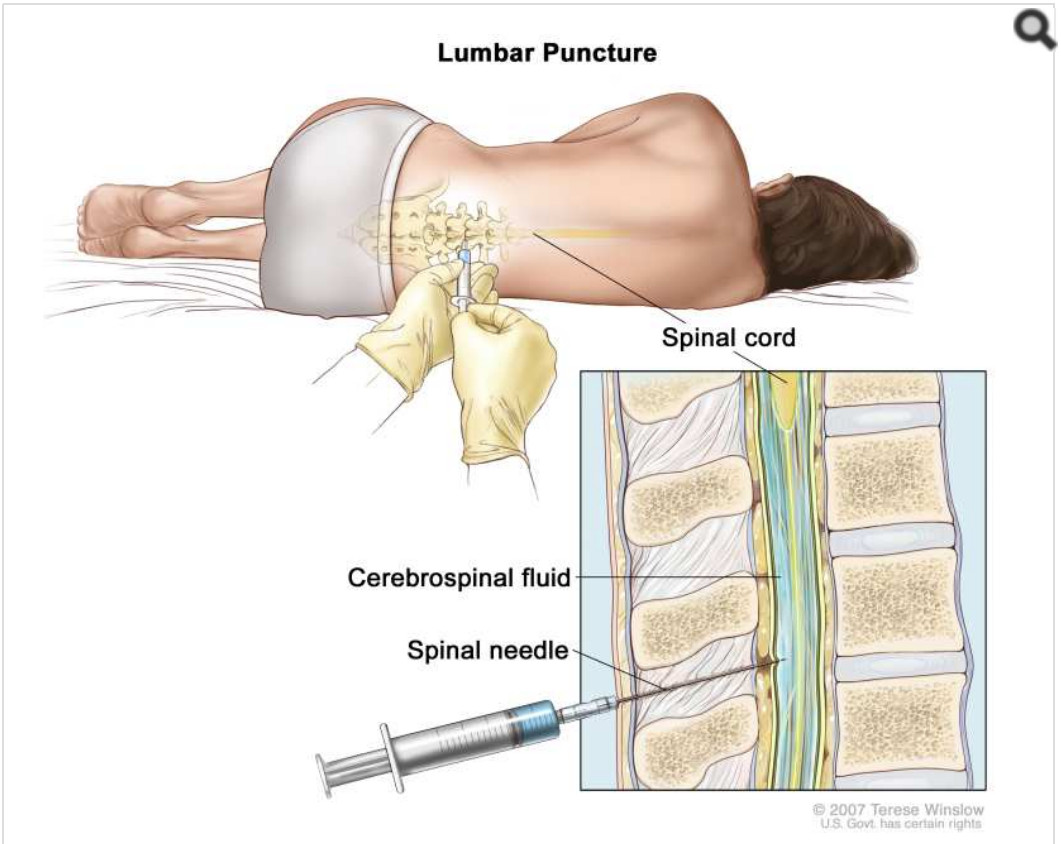
intermediate, and high risk. The risk groups are based on the amount of tumor remaining after surgery, the spread of cancer cells within the brain and spinal cord or to other parts of the body, where the tumor has formed, and the age of the child. The risk group is not used to plan treatment for all types of brain and spinal cord tumors.

The information from tests and procedures done to detect (find) childhood brain and spinal cord tumors is used to determine the tumor risk group.

After the tumor is removed in surgery, some of the tests used to detect childhood brain and spinal cord tumors are repeated to help determine the tumor risk group (see the General Information section). This is to find out how much tumor remains after surgery.

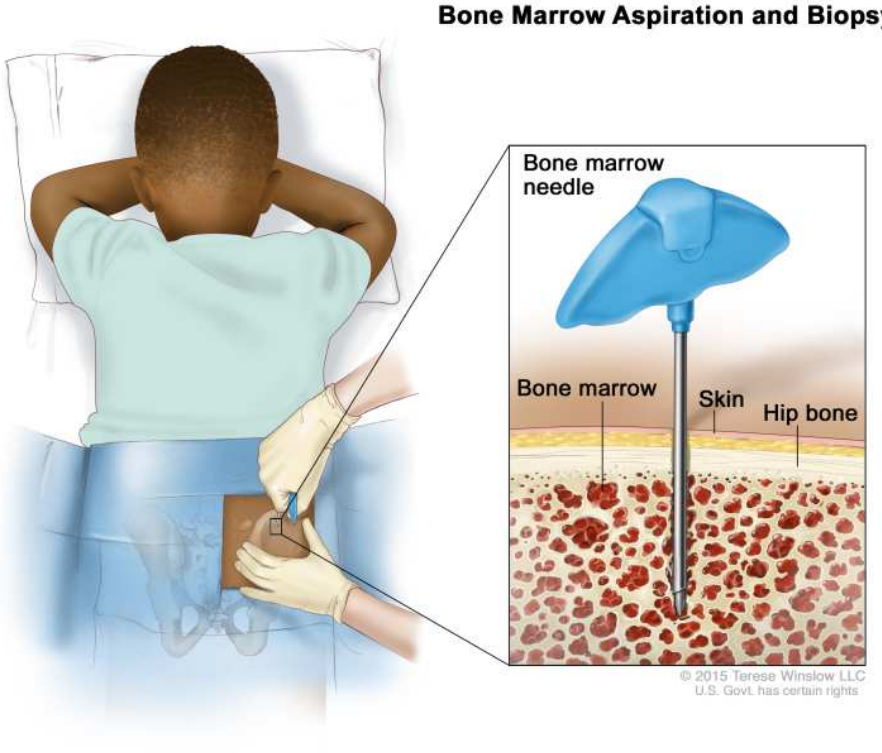
Other tests and procedures may be done to find out if cancer has spread:

- **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 the fluid. The sample of CSF is checked under a microscope for signs that the tumor has spread to the CSF. 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. A lumbar puncture is usually not used to stage childhood spinal cord tumors.



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.

- **Bone scan**: A procedure to check if there are rapidly dividing cells, such as cancer cells, in the bone. A very small amount of radioactive material is injected into a vein and travels through the bloodstream. The radioactive material collects in the bones with cancer and is detected by a scanner.
- **Bone marrow aspiration and biopsy**: The removal of bone marrow, blood, and a small piece of bone by inserting a hollow needle into the hipbone or breastbone. A pathologist views the bone marrow, blood, and bone under a microscope to look for signs of cancer.



Bone marrow aspiration and biopsy. After a small area of skin is numbed, a bone marrow needle is inserted into the child's hip bone. Samples of blood, bone, and bone marrow are removed for examination under a microscope.

Sometimes childhood brain and spinal cord tumors come back after treatment.

Childhood brain and spinal cord tumors may recur (come back) in the same place or in another part of the brain. Sometimes they come back in another part of the body. The tumor may come back many years after first being treated. Diagnostic and staging tests and procedures, including biopsy, may be done to make sure that the tumor has recurred.

Treatment Option Overview

Key Points for This Section

- There are different types of treatment for children with brain and spinal cord tumors.
- Children with brain or spinal cord tumors should have their treatment planned by a

team of health care providers who are experts in treating childhood brain and spinal cord tumors.

- Childhood brain and spinal cord tumors may cause signs or symptoms that begin before the cancer is diagnosed and continue for months or years.
- Three types of standard treatment are used:
  - Surgery
  - Radiation therapy
  - Chemotherapy
- New types of treatment are being tested in clinical trials.
- Treatment for childhood brain and spinal cord tumors 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 and spinal cord tumors.

Different types of treatment are available for children with brain and spinal cord tumors. 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. Clinical trials are taking place in many parts of the country. Some clinical trials are open only to patients who have not started treatment.

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

Treatment will be overseen by a pediatric oncologist, a doctor who specializes in treating children with cancer. The pediatric oncologist works with other 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.
- Neurologist.
- Neuro-oncologist.
- Neuropathologist.

- Neuroradiologist.
- Radiation oncologist.
- Endocrinologist.
- Psychologist.
- Ophthalmologist.
- Rehabilitation specialist.
- Social worker.
- Nurse specialist.

Childhood brain and spinal cord tumors may cause signs or symptoms that begin before the cancer is diagnosed and continue for months or years.

Childhood brain and spinal cord tumors may cause signs or symptoms that continue for months or years. Signs or symptoms caused by the tumor may begin before diagnosis. Signs or symptoms caused by treatment may begin during or right after treatment.

Three types of standard treatment are used:

Surgery

Surgery may be used to diagnose and treat childhood brain and spinal cord tumors. See the General Information section of this summary.

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.

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). Although most cannot, some chemotherapy drugs can cross the blood-brain barrier and reach tumor cells in the brain. When chemotherapy is placed directly in the cerebrospinal fluid, it is called intrathecal chemotherapy. The way the chemotherapy is given depends on the type and grade of tumor and where it is in the brain or spinal cord.

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.

High-dose chemotherapy with stem cell transplant

High doses of chemotherapy are given to kill cancer cells. Healthy cells, including blood-forming cells, are also destroyed by the cancer treatment. Stem cell transplant is a treatment to replace the blood-forming cells. Stem cells (immature blood cells) are removed from the blood or bone marrow of the patient or a donor and are frozen and stored. After the patient completes chemotherapy, the stored stem cells are thawed and given back to the patient through an infusion. These reinfused stem cells grow into (and restore) the body's blood cells.

Treatment for childhood brain and spinal cord tumors 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.
- 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.

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.

Treatment of Newly Diagnosed and Recurrent Childhood Brain Tumors

The brain is made of different kinds of cells. Childhood brain tumors are grouped and treated based on the type of cell the cancer formed in and where the tumor began growing in the CNS. Some types of tumors are divided into subtypes based on how the tumor looks under a microscope and whether it has certain gene changes. See Table 1 for a list of tumor types and staging and treatment information for newly diagnosed and recurrent childhood brain tumors.

Table 1. Newly Diagnosed or Recurrent Tumor Type and Its Related PDQ Treatment Summary

Tumor Type	Tumor Subtype	Related PDQ Treatment Summary
Astrocytomas and Other Tumors of Glial Origin		
– *Low-Grade Astrocytomas*	Angiocentric glioma	See Childhood Astrocytomas Treatment for information on low-grade astrocytomas.
	Choroid glioma of the third ventricle	
	Diffuse astrocytoma, *IDH*-mutant, *IDH*-wild type, or NOS	
	Pilocytic astrocytoma	
	Pleomorphic xanthoastrocytoma	
	Subependymal giant cell astrocytoma	
– *High-Grade Astrocytomas*	Anaplastic astrocytoma, *IDH*-mutant or *IDH*-wild type	See Childhood Astrocytomas Treatment for information on high-grade astrocytomas.
	Anaplastic pleomorphic xanthoastrocytoma	
	Diffuse midline glioma, H3 K27M-mutant	
	Glioblastoma, *IDH*-mutant	
	Glioblastoma, *IDH*-wildtype	

Tumor Type	Tumor Subtype	Related PDQ Treatment Summary
– *Other Astrocytomas or Gliomas*	Astroblastoma	See Childhood Astrocytomas Treatment for information on other astrocytomas or gliomas.
	Pilomyxoid astrocytoma	
Brain Stem Glioma		
	Diffuse intrinsic pontine gliomas, H3 K27M-mutant	See Childhood Brain Stem Glioma Treatment.
	Focal or low-grade brain stem gliomas	
Central Nervous System Embryonal Tumors		
– *Medulloblastoma*		See Childhood Medulloblastoma and Other Central Nervous System Embryonal Tumors Treatment for information on medulloblastoma and nonmedulloblastomas.
– *Nonmedulloblastomas*	Central nervous system ganglioneuroblastoma	
	Central nervous system neuroblastoma	
	Embryonal tumor with multilayered rosettes, *C19MC*-altered or NOS	
	Medulloepithelioma	
– *Central Nervous System Atypical Teratoid/Rhabdoid Tumor*		See Childhood Central Nervous System Atypical Teratoid/Rhabdoid Tumor Treatment.
Tumors of the Pineal Region	Pineoblastoma	See Childhood Medulloblastoma and Other Central Nervous System Embryonal Tumors Treatment.
Central Nervous System Germ Cell Tumors		
– *Germinomas*		See Childhood Central Nervous System Germ Cell Tumors Treatment.
– *Teratomas*	Immature teratomas	
	Mature teratomas	
	Teratomas with malignant transformation	
– *Non-germinomatous Germ Cell Tumors*	Choriocarcinoma	
	Embryonal carcinoma	
	Mixed germ cell tumors	
	Yolk sac tumor	
Craniopharyngioma		See Childhood Craniopharyngioma Treatment.

Tumor Type	Tumor Subtype	Related PDQ Treatment Summary
Ependymoma		
	Subependymoma (WHO grade I)	See Childhood Ependymoma Treatment.
	Myxopapillary ependymoma (WHO grade I)	
	Ependymoma (WHO grade II)	
	RELA fusion-positive ependymoma (WHO grade II or grade III)	
	Anaplastic ependymoma (WHO grade III)	
Tumors of the Choroid Plexus		

NOS = not otherwise specified; WHO = World Health Organization.

Treatment of Newly Diagnosed and Recurrent Childhood Spinal Cord Tumors

Tumors of many different cell types may form in the spinal cord. Low-grade spinal cord tumors usually do not spread. High-grade spinal cord tumors may spread to other places in the spinal cord or to the brain. See the following PDQ summaries for more information on staging and treatment of newly diagnosed and recurrent childhood spinal cord tumors:

- Childhood Astrocytomas Treatment
- Childhood Medulloblastoma and Other Central Nervous System Embryonal Tumors Treatment
- Childhood Ependymoma Treatment

To Learn More About Childhood Brain and Spinal Cord Tumors

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

- Pediatric Brain Tumor Consortium (PBTC)

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

Physician Data Query (PDQ) is the National Cancer Institute's (NCI's) comprehensive cancer information database. The PDQ database contains summaries of the latest published information on cancer prevention, detection, genetics, treatment, supportive care, and complementary and alternative medicine. Most summaries come in two versions. The health professional versions have detailed information written in technical language. The patient versions are written in easy-to-understand, nontechnical language. Both versions have cancer information that is accurate and up to date and most versions are also available in Spanish.

PDQ is a service of the NCI. The NCI is part of the National Institutes of Health (NIH). NIH is the federal government’s center of biomedical research. The PDQ summaries are based on an independent review of the medical literature. They are not policy statements of the NCI or the NIH.

Purpose of This Summary

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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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Contact Us

More information about contacting us or receiving help with the Cancer.gov website can be found on our Contact Us for Help page. Questions can also be submitted to Cancer.gov through the website's E-mail Us.

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