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# Childhood Medulloblastoma and Other Central Nervous System Embryonal Tumors Treatment (PDQ®)

Patient Version

#### Authors

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This PDQ cancer information summary has current information about the treatment of childhood medulloblastoma and other central nervous system embryonal 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 Medulloblastoma and Other Central Nervous System Embryonal Tumors

#### **Key Points for This Section**

- Medulloblastoma and other central nervous system (CNS) embryonal tumors may begin in embryonic (fetal) cells that remain in the brain after birth.
- There are different types of CNS embryonal tumors.
- Pineoblastomas form in cells of the pineal gland.
- Certain genetic conditions increase the risk of childhood medulloblastoma and other CNS embryonal tumors.
- Signs and symptoms of childhood medulloblastoma and other CNS embryonal tumors depend on the child's age and where the tumor is.
- Tests that examine the brain and spinal cord are used to diagnose childhood medulloblastoma and other CNS embryonal tumors.
- A biopsy may be done to be sure of the diagnosis of medulloblastoma and other CNS embryonal tumors.
- Certain factors affect prognosis (chance of recovery) and treatment options.

# Medulloblastoma and other central nervous system (CNS) embryonal tumors may begin in embryonic (fetal) cells that remain in the brain after birth.

These tumors tend to spread through the cerebrospinal fluid (CSF) to other parts of the brain and spinal cord.

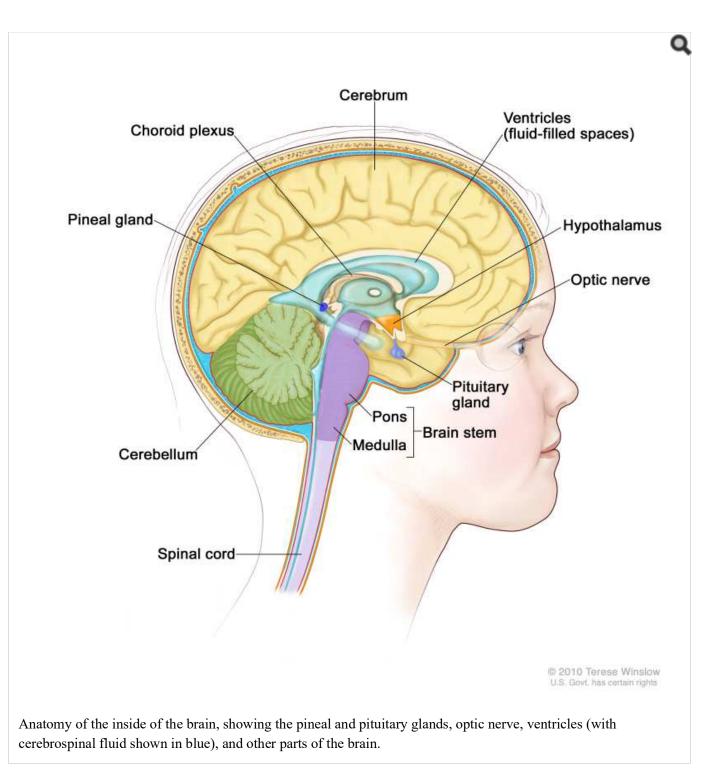
<u>Medulloblastoma</u> and other <u>CNS</u> embryonal tumors may be malignant (cancer) or <u>benign</u> (not cancer). Most of the tumors in children are malignant. Malignant <u>brain tumors</u> are likely to grow quickly and spread into other parts of the brain. When a tumor grows into or presses on an area of the brain, it may stop that part of the brain from working the way it should. Benign brain tumors grow and press on nearby areas of the brain. They rarely spread to other parts of the brain. Both benign and malignant brain tumors can cause signs or symptoms and need treatment.

Although cancer is rare in children, brain tumors are the second most common type of childhood cancer, after leukemia. This summary is about the treatment of primary brain tumors (tumors that begin in the brain). For information about the different types of brain and spinal cord tumors, see the PDQ summary on Childhood Brain and Spinal Cord Tumors Treatment Overview.

Brain tumors occur in both children and adults. Treatment for adults may be different from treatment for children. See the PDQ summary on <u>Adult Central Nervous System Tumors Treatment</u> for more information on the treatment of adults.

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

There are different types of CNS embryonal tumors.



The different types of CNS embryonal tumors include:

#### Medulloblastomas

Most CNS embryonal tumors are medulloblastomas. Medulloblastomas are fast-growing tumors that form in brain cells in the cerebellum. The cerebellum is at the lower back part of the brain between the cerebrum and the brain stem. The cerebellum controls movement, balance, and posture. Medulloblastomas sometimes spread to the bone, bone marrow, lung, or other parts of the body, but this is rare.

#### Nonmedulloblastoma embryonal tumors

Nonmedulloblastoma embryonal tumors are fast-growing tumors that usually form in brain cells in the

cerebrum. The cerebrum is at the top of the head and is the largest part of the brain. The cerebrum controls thinking, learning, problem-solving, emotions, speech, reading, writing, and voluntary movement. Nonmedulloblastoma embryonal tumors may also form in the brain stem or spinal cord. These tumors sometimes spread to the bone, bone marrow, lung, or other parts of the body, but this is rare.

There are six types of nonmedulloblastoma embryonal tumors:

### • Embryonal tumors with multilayered rosettes

Embryonal tumors with multilayered rosettes (ETMR) are rare tumors that form in the brain and spinal cord. ETMR most commonly occur in young children and are fast-growing tumors.

#### • Medulloepitheliomas

Medulloepitheliomas are fast-growing tumors that usually form in the brain, spinal cord, or <u>nerves</u> just outside the spinal column. They occur most often in infants and young children.

### • CNS neuroblastomas

CNS <u>neuroblastomas</u> are a very rare type of neuroblastoma that form in the nerve <u>tissue</u> of the cerebrum or the layers of tissue that cover the brain and spinal cord. CNS neuroblastomas may be large and spread to other parts of the brain or spinal cord.

### • CNS ganglioneuroblastomas

CNS ganglioneuroblastomas are rare tumors that form in nerve tissue of the brain and spinal cord. They may form in one area and be fast growing or form in more than one area and be slow growing.

### • CNS embryonal tumor, NOS

Central nervous system embryonal tumor, not otherwise specified (NOS) was previously called primitive neuroectodermal tumors (PNET). These are rare tumors that form in the brain and spinal cord. CNS embryonal tumor, NOS most commonly occurs in young children.

### • CNS embryonal tumor with rhabdoid features

CNS embryonal tumor with rhabdoid features is a rare tumor that forms in the brain and spinal cord. These tumors are similar to CNS atypical teratoid/rhabdoid tumors but lack certain gene changes.

CNS atypical teratoid/rhabdoid tumor is also a type of embryonal tumor, but it is treated differently than other childhood CNS embryonal tumors. See the PDQ summary on <u>Childhood Central Nervous System Atypical</u> Teratoid/Rhabdoid Tumor Treatment for more information.

### Pineoblastomas form in cells of the pineal gland.

The pineal gland is a tiny organ in the center of the brain. The gland makes melatonin, a substance that helps control our sleep cycle.

Pineoblastomas form in cells of the pineal gland and are usually malignant. Pineoblastomas are fast-growing tumors with cells that look very different from normal pineal gland cells. Pineoblastomas are not a type of CNS embryonal tumor but treatment for them is a lot like treatment for CNS embryonal tumors.

Pineoblastoma is linked with inherited changes in the *retinoblastoma* (*RB1*) gene. A child with the inherited form of retinoblastoma (cancer that forms in the tissues of the retina) has an increased risk of pineoblastoma. When retinoblastoma forms at the same time as a tumor in or near the pineal gland, it is called trilateral retinoblastoma. <u>MRI</u> (magnetic resonance imaging) testing in children with retinoblastoma may detect pineoblastoma at an early stage when it can be treated successfully.

# Certain genetic conditions increase the risk of childhood medulloblastoma and other CNS embryonal tumors.

Anything that increases the risk of getting a disease is called a <u>risk factor</u>. 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.

Risk factors for medulloblastoma and other CNS embryonal tumors include having the following inherited diseases:

- Turcot syndrome.
- Rubinstein-Taybi syndrome.
- Nevoid basal cell carcinoma (Gorlin) syndrome.
- Li-Fraumeni syndrome.
- Fanconi anemia.

Children with certain gene changes or a <u>family history</u> of cancers linked to changes in the *BRCA* gene may be considered for genetic testing. Although rare, this is to check whether the child has a cancer predisposition <u>syndrome</u> that places the child at risk for other diseases or types of cancer.

In most cases, the cause of medulloblastoma and other CNS embryonal tumors is not known.

# Signs and symptoms of childhood medulloblastoma and other CNS embryonal tumors depend on the child's age and where the tumor is.

These and other signs and symptoms may be caused by childhood medulloblastoma and other CNS embryonal tumors or other conditions. Check with your child's doctor if your child has any of the following:

- Loss of balance, trouble walking, lack of coordination, or slow speech.
- Headache, especially in the morning, or headache that goes away after vomiting.
- General weakness or weakness on one side of the face.
- Unusual sleepiness or change in energy level.
- Seizures.
- Double vision or other eye problems.
- Nausea and vomiting.

Infants and young children with these tumors may be irritable or grow slowly. Also they may not eat well or meet developmental milestones such as sitting, walking, and talking in sentences.

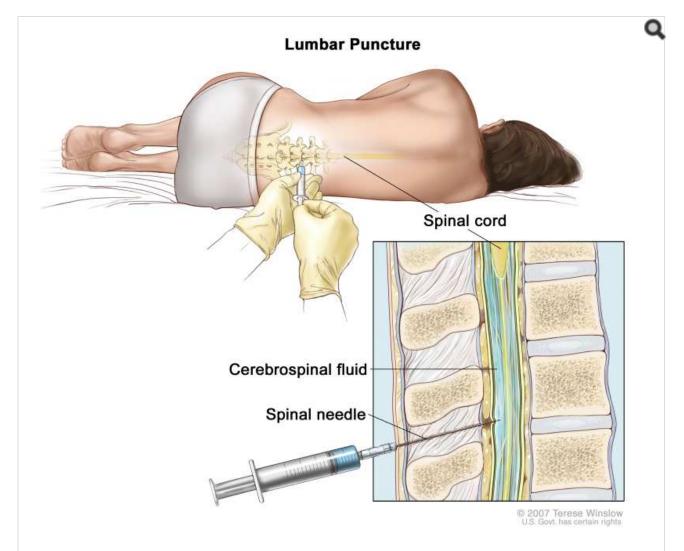
# Tests that examine the brain and spinal cord are used to diagnose childhood medulloblastoma and other CNS embryonal tumors.

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 patient'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) of the brain and spinal cord 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. 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). Sometimes magnetic resonance spectroscopy (MRS) is done during the MRI scan to look at the chemicals in brain tissue.
- <u>CT scan</u> (CAT scan): A procedure that makes a series of detailed pictures of areas inside the body, taken from different angles. The pictures are made by a computer linked to an <u>x-ray</u> machine. A dye may be injected into a vein or swallowed to help the organs or tissues show up more clearly. This procedure is also called computed tomography, computerized tomography, or computerized axial tomography.
- 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 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.

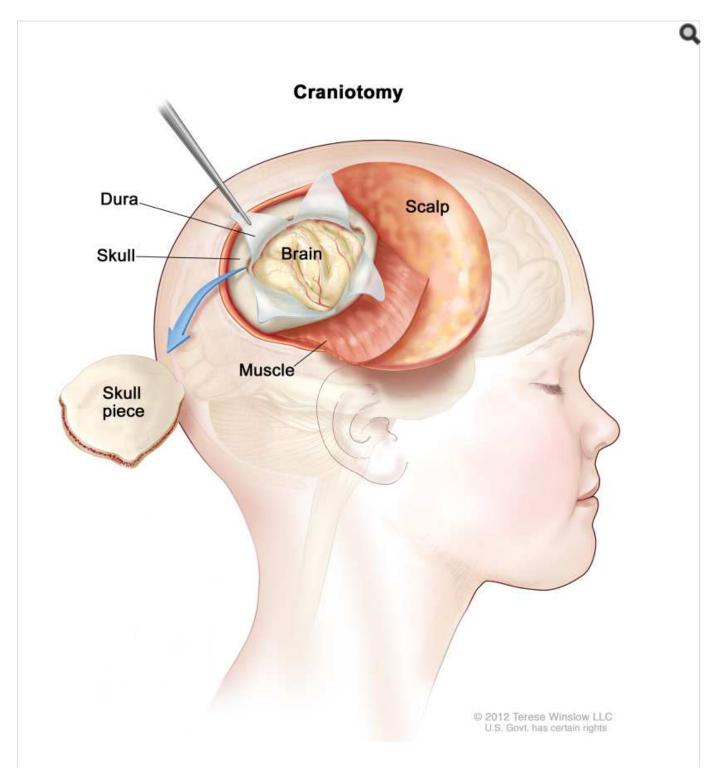


Lumbar puncture. A patient lies in a curled position on a table. After a small area on the lower back is numbed, a spinal needle (a long, thin needle) is inserted into the lower part of the spinal column to remove

cerebrospinal fluid (CSF, shown in blue). The fluid may be sent to a laboratory for testing.

# A biopsy may be done to be sure of the diagnosis of medulloblastoma and other CNS embryonal tumors.

If doctors think your child may have a medulloblastoma or other CNS embryonal tumor, a <u>biopsy</u> may be done. For brain tumors, the biopsy is done by removing part of the <u>skull</u> and using a needle to remove a sample of tissue. Sometimes, a computer-guided needle is used to remove the tissue sample. A <u>pathologist</u> 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 piece of skull is usually put back in place after the procedure.



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 <u>enzyme</u> 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 <u>diagnose</u> 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:

- The type of tumor and where it is in the brain.
- Whether the cancer has spread within the brain and spinal cord when the tumor is found.
- The age of the child when the tumor is found.
- How much of the tumor remains after surgery.
- Whether there are certain changes in the chromosomes, genes, or brain cells.
- Whether the tumor has just been diagnosed or has recurred (come back).

# Staging Childhood Medulloblastoma and Other Central Nervous System Embryonal Tumors

#### **Key Points for This Section**

- Treatment of childhood medulloblastoma and other central nervous system (CNS) embryonal tumors depends on the type of tumor and the child's age.
- Treatment of medulloblastoma in children older than 3 years also depends on whether the tumor is average risk or high risk.
  - Average risk (child is older than 3 years of age)
  - High risk (child is older than 3 years of age)
- The information from tests and procedures done to diagnose childhood medulloblastoma and other CNS embryonal tumors is used to plan cancer treatment.
- Sometimes childhood medulloblastoma and other central nervous system embryonal tumors come back after treatment.

# Treatment of childhood medulloblastoma and other central nervous system (CNS) embryonal tumors depends on the type of tumor and the child's age.

<u>Staging</u> is the process used to find out how much <u>cancer</u> 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 medulloblastoma and other central nervous system (CNS) embryonal tumors. Instead, treatment depends on the type of tumor and the child's age (3 years and younger or older than 3 years).

# Treatment of medulloblastoma in children older than 3 years also depends on whether the tumor is average risk or high risk.

#### Average risk (child is older than 3 years of age)

Medulloblastomas are called average risk when all of the following are true:

- The tumor was completely removed by surgery or there was only a very small amount remaining.
- The cancer has not spread to other parts of the body.

#### High risk (child is older than 3 years of age)

Medulloblastomas are called high risk if any of the following are true:

- Some of the tumor was not removed by surgery.
- The cancer has spread to other parts of the brain or spinal cord or to other parts of the body.

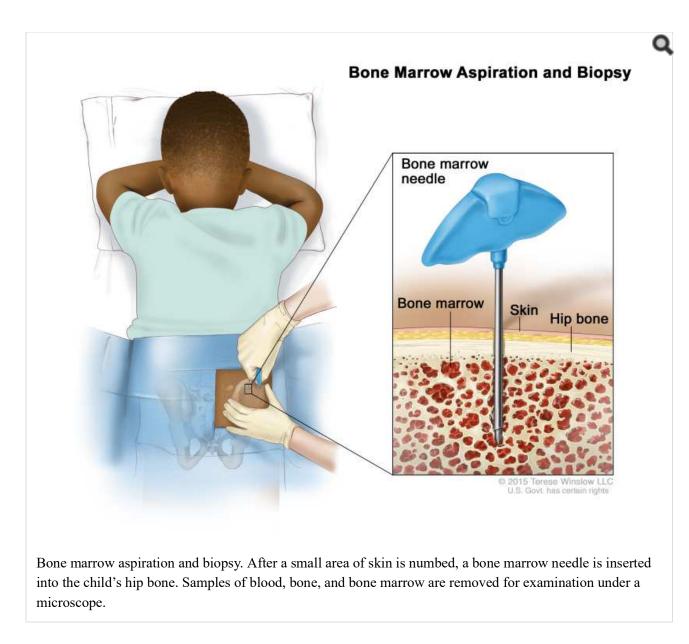
In general, cancer is more likely to recur (come back) in patients with a high-risk tumor.

# The information from tests and procedures done to diagnose childhood medulloblastoma and other CNS embryonal tumors is used to plan cancer treatment.

Some of the tests used to detect childhood medulloblastoma and other CNS embryonal tumors are repeated after surgery to remove the tumor. (See the <u>General Information</u> section.) This is to find out how much tumor remains after surgery.

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

• **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. A bone marrow aspiration and biopsy is only done when there are signs the cancer has spread to the bone marrow.



- **Bone scan**: A procedure to check if there are rapidly dividing <u>cells</u>, 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. A bone scan is only done when there are signs or symptoms that the cancer has spread to the bone.
- Chest x-ray: An x-ray of the organs and bones inside the chest. An x-ray is a type of energy beam that can go through the body and onto film, making a picture of areas inside the body. A chest x-ray is only done when there are signs or symptoms that the cancer has spread to the chest.
- 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 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.

# Sometimes childhood medulloblastoma and other central nervous system embryonal tumors come back after treatment.

Childhood medulloblastoma and other CNS embryonal tumors most often recur (come back) within 3 years after

treatment but may come back many years later. <u>Recurrent</u> childhood medulloblastoma and other CNS embryonal tumors may come back in the same place as the original tumor and/or in a different place in the brain or <u>spinal cord</u>. Medulloblastoma and other CNS embryonal tumors rarely spread to other parts of the body.

## **Treatment Option Overview**

#### **Key Points for This Section**

- There are different types of treatment for children who have medulloblastoma and other central nervous system (CNS) embryonal tumors.
- Children who have medulloblastoma and other CNS embryonal tumors should have their treatment planned by a team of health care providers who are experts in treating brain tumors in children.
- Childhood brain tumors may cause signs or symptoms that begin before the cancer is diagnosed and continue for months or years.
- Five types of treatment are used:
  - Surgery
  - Radiation therapy
  - Chemotherapy
  - High-dose chemotherapy with stem cell rescue
  - Targeted therapy
- New types of treatment are being tested in clinical trials.
- Treatment for childhood medulloblastoma and other central nervous system embryonal 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 who have medulloblastoma and other central nervous system (CNS) embryonal tumors.

Different types of treatment are available for children with <u>medulloblastoma</u> and other <u>central nervous system</u> (CNS) <u>embryonal tumors</u>. Some treatments are <u>standard</u> (the currently used treatment), and some are being tested in <u>clinical</u> trials. A treatment clinical trial is a <u>research study</u> meant to help improve current treatments or obtain information on new treatments for patients with <u>cancer</u>. 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 who have medulloblastoma and other CNS embryonal tumors should have their treatment planned by a team of health care providers who are experts in treating brain tumors in children.

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

- Pediatrician.
- <u>Neurosurgeon</u>.
- Neurologist.
- Neuropathologist.
- Neuroradiologist.
- Rehabilitation specialist.
- Radiation oncologist.
- Psychologist.

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

Signs or symptoms caused by the tumor may begin before the cancer is diagnosed and continue for months or years. It is important to talk with your child's doctors about signs or symptoms caused by the tumor that may continue after treatment.

### Five types of treatment are used:

#### Surgery

<u>Surgery</u> is used to diagnose and treat childhood medulloblastoma and other CNS embryonal tumors as described in the General Information section of this summary.

After the doctor removes all the cancer that can be seen at the time of the surgery, some patients may be given chemotherapy, radiation therapy, or both after surgery to kill any cancer <u>cells</u> that are left. Treatment given after the 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 <u>x-rays</u> or other types of <u>radiation</u> 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 <u>tissue</u>. These types of radiation therapy include the following:

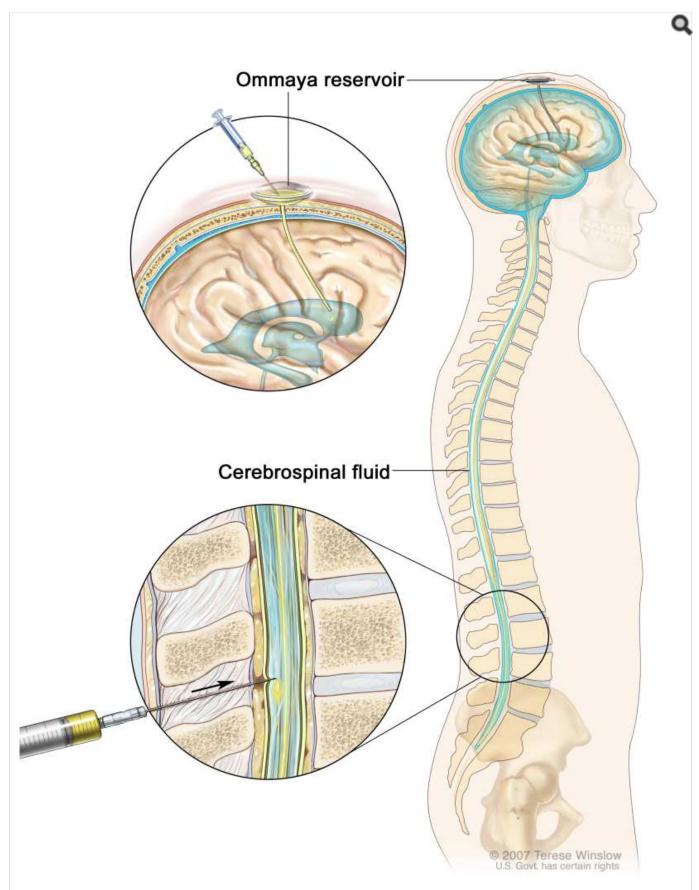
- <u>Conformal radiation therapy</u>: Conformal radiation therapy is a type of external radiation therapy that uses a computer to make a <u>3-dimensional</u> (3-D) picture of the tumor and shapes the radiation beams to fit the tumor. This allows a high dose of radiation to reach the tumor and causes less damage to nearby healthy tissue.
- Stereotactic radiation therapy: Stereotactic radiation therapy is a type of external radiation therapy. A rigid head frame is attached to the <u>skull</u> to keep the head still during the radiation treatment. A machine aims radiation directly at the tumor, causing less damage to nearby healthy tissue. The total dose of radiation is divided into several smaller doses given over several days. This procedure is also called stereotactic external-beam radiation therapy and stereotaxic radiation therapy.

Because radiation therapy can affect growth and brain development in young children, especially children who are 3 years old or younger, chemotherapy may be given to delay or reduce the need for radiation therapy.

Radiation therapy to the brain can also affect growth and development in children older than three. For this reason, clinical trials are studying new ways of giving radiation that may have fewer side effects than standard methods.

#### Chemotherapy

Chemotherapy is a cancer treatment that uses <u>drugs</u> 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 <u>injected</u> into a <u>vein</u> 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 <u>blood-brain barrier</u> and reach tumor cells in the brain. When chemotherapy is placed directly into the <u>cerebrospinal fluid</u>, it is called <u>intrathecal chemotherapy</u>. Combination chemotherapy is treatment using more than one anticancer drug. The way the chemotherapy is given depends on the type of cancer being treated.



Intrathecal chemotherapy. Anticancer drugs are injected into the intrathecal space, which is the space that holds the cerebrospinal fluid (CSF, shown in blue). There are two different ways to do this. One way, shown in the top part of the figure, is to inject the drugs into an Ommaya reservoir (a dome-shaped container that is placed under the scalp during surgery; it holds the drugs as they flow through a small tube into the brain). The other way, shown in

the bottom part of the figure, is to inject the drugs directly into the CSF in the lower part of the spinal column, after a small area on the lower back is numbed.

#### High-dose chemotherapy with stem cell rescue

High doses of chemotherapy are given to kill cancer cells. Healthy cells, including <u>blood</u>-forming cells, are also destroyed by the cancer treatment. <u>Stem cell transplant</u> is a treatment to replace the blood-forming cells. <u>Stem cells</u> (immature blood cells) are removed from the blood or <u>bone marrow</u> of the patient or a <u>donor</u> 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.

#### **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.

Signal transduction inhibitors: This treatment blocks signals that are passed from one molecule to another inside a cell. Blocking these signals may kill cancer cells. <u>Vismodegib</u> is a type of signal transduction inhibitor that may be used to treat recurrent medulloblastoma in children who have finished growing.

Targeted therapy is being studied for the treatment of childhood medulloblastoma and other CNS embryonal tumors that have recurred (come back).

### New types of treatment are being tested in clinical trials.

Information about clinical trials is available from the NCI website.

# Treatment for childhood medulloblastoma and other central nervous system embryonal 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 <u>late effects</u>. Late effects of cancer treatment may include the following:

- Physical problems affect the following:
  - Bone and muscle growth and development.
  - Thyroid, heart, or hearing function.
- Changes in mood, feelings, thinking, learning, or memory.
- Second cancers (new types of cancer), such as thyroid or other brain tumors.

Children diagnosed with medulloblastoma may have certain problems after surgery or radiation therapy such as changes in the ability to think, learn, and pay attention. Also, cerebellar mutism syndrome may occur after surgery. Signs of this syndrome include the following:

- Delayed ability to speak.
- Trouble swallowing and eating.
- Loss of balance, trouble walking, and worsening handwriting.
- Loss of muscle tone.

• Mood swings and changes in personality.

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 and the types of symptoms to expect after cancer treatment has ended. (See the <u>PDQ</u> 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 <u>clinical trial</u> 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 <u>clinical trials search</u> 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 <u>diagnose</u> the cancer or to find out the <u>stage</u> of the cancer may be repeated. (See the <u>General Information</u> section for a list of tests.) 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. This is sometimes called re-staging.

Some of the <u>imaging tests</u> will continue to be done from time to time after treatment has ended. The results of these tests can show if your child's <u>condition</u> has changed or if the brain tumor has recurred (come back). If the imaging tests show abnormal tissue in the brain, a biopsy may also be done to find out if the tissue is made up of dead tumor cells or if new cancer cells are growing. These tests are sometimes called follow-up tests or check-ups.

### **Treatment of Childhood Medulloblastoma**

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

#### Children aged 3 years and younger

Standard treatment of newly diagnosed medulloblastoma in children aged 3 years and younger includes the following:

• Surgery to remove as much of the tumor as possible, followed by chemotherapy.

Other treatments that may be given after surgery include the following:

• High-dose chemotherapy with stem cell rescue.

• Chemotherapy with or without radiation therapy to the area where the tumor was removed.

#### Children older than 3 years with average-risk medulloblastoma

Standard treatment of newly diagnosed average-risk medulloblastoma in children older than 3 years includes the following:

- Surgery to remove as much of the tumor as possible. This is followed by radiation therapy to the brain and spinal cord. Chemotherapy may also be given during and after radiation therapy.
- Surgery to remove the tumor, radiation therapy, and high-dose chemotherapy with stem cell rescue.
- A clinical trial of reduced doses of radiation therapy to the brain and spinal cord and chemotherapy for patients with a certain type of medulloblastoma.

#### Children older than 3 years with high-risk medulloblastoma

Standard treatment of newly diagnosed high-risk medulloblastoma in children older than 3 years includes the following:

- Surgery to remove as much of the tumor as possible. This is followed by a larger dose of radiation therapy to the brain and spinal cord than the dose given for average-risk medulloblastoma. Chemotherapy is also given during and after radiation therapy.
- Surgery to remove the tumor, radiation therapy, and high-dose chemotherapy with stem cell rescue.

### Treatment of Childhood Nonmedulloblastoma Embryonal Tumors

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

# Children aged 3 years and younger with nonmedulloblastoma, nonmedulloepithelioma embryonal tumors, embryonal tumors with multilayered rosettes, or medulloepithelioma

Standard treatment of newly diagnosed nonmedulloblastoma, nonmedulloepithelioma embryonal tumors, embryonal tumors with multilayered rosettes, or medulloepithelioma in children 3 years or younger includes the following:

• Surgery to remove as much of the tumor as possible, followed by chemotherapy.

Other treatments that may be given after surgery include the following:

- High-dose chemotherapy with stem cell rescue.
- Chemotherapy and radiation therapy to the area where the tumor was removed.

#### Children older than 3 years with nonmedulloblastoma, nonmedulloepithelioma embryonal tumors

Standard treatment of newly diagnosed nonmedulloblastoma, nonmedulloepithelioma embryonal tumors in children older than 3 years includes the following:

• Surgery to remove as much of the tumor as possible. This is followed by radiation therapy to the brain and spinal cord. Chemotherapy is also given during and after radiation therapy.

#### Children older than 3 years with embryonal tumors with multilayered rosettes or medulloepithelioma

Standard treatment of newly diagnosed embryonal tumor with multilayered rosettes (ETMR) or medulloepithelioma includes the following:

- Surgery to remove as much of the tumor as possible. This is followed by radiation therapy to the brain and spinal cord. Chemotherapy is also given during and after radiation therapy.
- Surgery to remove the tumor, radiation therapy, and high-dose chemotherapy with stem cell rescue.

# **Treatment of Childhood Pineoblastoma**

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

#### Children aged 3 years and younger

Standard treatment of newly diagnosed pineoblastoma in children aged 3 years and younger includes the following:

- Biopsy to diagnose pineoblastoma followed by chemotherapy.
- If the tumor responds to chemotherapy, radiation therapy is given when the child is older.
- High-dose chemotherapy with stem cell rescue.

#### Children older than 3 years

Standard treatment of newly diagnosed pineoblastoma in children older than 3 years includes the following:

- <u>Surgery</u> to remove the tumor. The tumor usually cannot be completely removed because of where it is in the brain. Surgery is often followed by radiation therapy to the brain and spinal cord and chemotherapy.
- A clinical trial of high-dose chemotherapy after radiation therapy and stem cell rescue.
- A clinical trial of chemotherapy during radiation therapy.

### Treatment of Recurrent Childhood Medulloblastoma and Other Central Nervous System Embryonal Tumors

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

Treatment for recurrent childhood medulloblastoma and other CNS embryonal tumors may include the following:

- Surgery to remove as much of the tumor as possible.
- For children who previously received <u>radiation therapy</u> and <u>chemotherapy</u>, treatment may include repeat radiation at the site where the <u>cancer</u> started and where the tumor has spread. <u>Stereotactic radiation therapy</u> and/or chemotherapy may also be used.
- For infants and young children who previously received chemotherapy only and have a local recurrence, treatment may be chemotherapy with radiation therapy to the tumor and the area close to it. Surgery to remove the tumor may also be done.
- For patients who previously received radiation therapy, high-dose chemotherapy and stem cell rescue may be used. It is not known whether this treatment improves survival.
- Targeted therapy with a signal transduction inhibitor (vismodegib) for patients whose cancer has certain changes in the genes.
- 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.

Use our <u>clinical trial search</u> to find NCI-supported cancer clinical trials that are accepting patients. You can search for trials based on the type of cancer, the age of the patient, and where the trials are being done. <u>General information</u> about

clinical trials is also available.

### To Learn More About Childhood Medulloblastoma and Other Central Nervous System Embryonal Tumors

For more information about childhood medulloblastoma and other central nervous system embryonal tumor, see the following:

- Pediatric Brain Tumor Consortium (PBTC)
- Targeted Cancer Therapies
- Computed Tomography (CT) Scans and Cancer

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**

This PDQ cancer information summary has current information about the treatment of childhood medulloblastoma and other central nervous system embryonal 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.

#### **Reviewers and Updates**

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 ("Updated") 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.

#### **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 <u>Contact Us</u> for Help page. Questions can also be submitted to Cancer.gov through the website's <u>E-mail Us</u>.

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