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Childhood Craniopharyngioma Treatment (PDQ®)

Patient Version

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This PDQ cancer information summary has current information about the treatment of childhood craniopharyngioma. 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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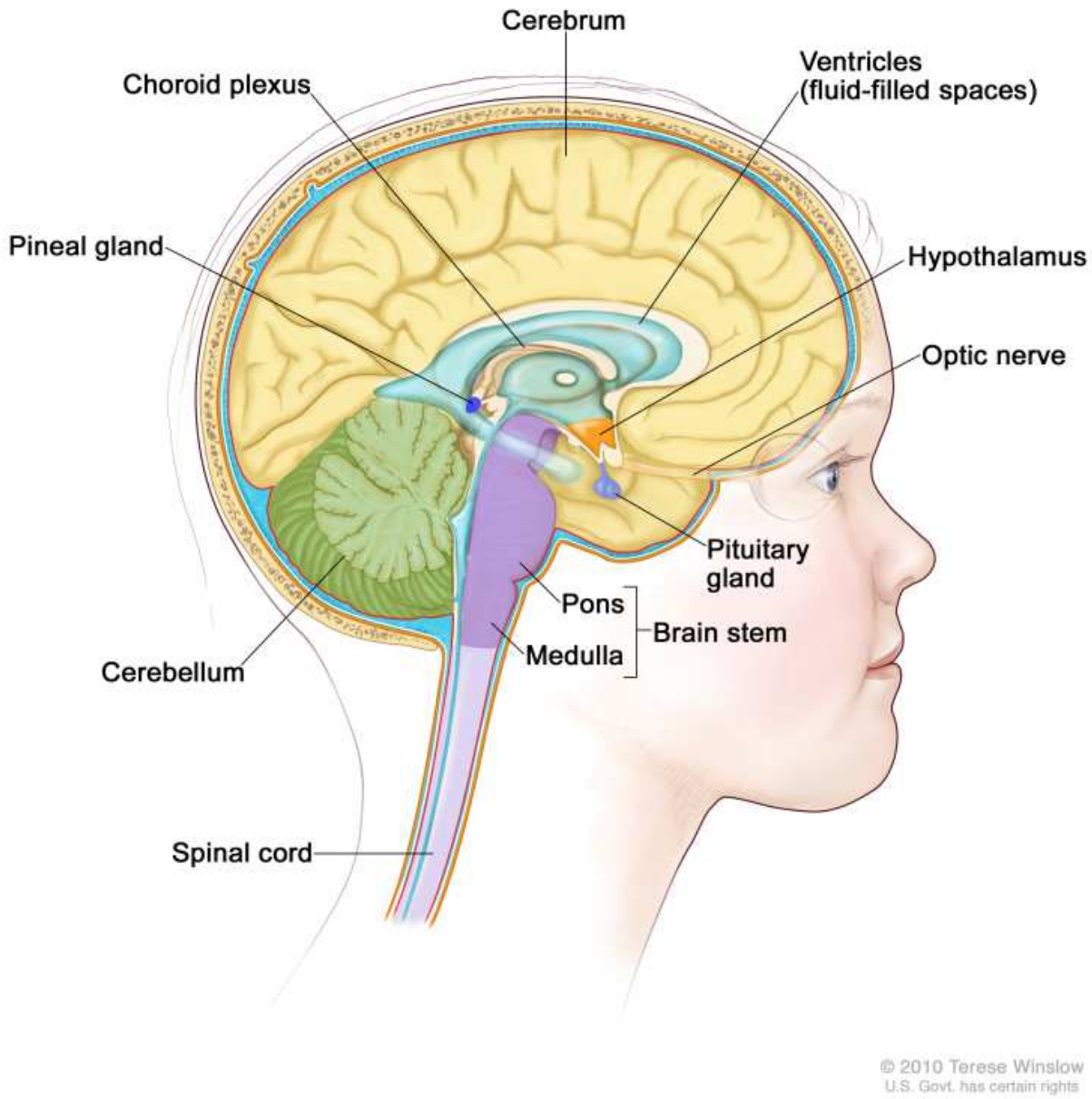
General Information About Childhood Craniopharyngioma

Key Points for This Section

- Childhood craniopharyngiomas are benign brain tumors found near the pituitary gland.
- There are no known risk factors for childhood craniopharyngioma.
- Signs of childhood craniopharyngioma include vision changes and slow growth.
- Tests that examine the brain, vision, and hormone levels are used to detect (find) childhood craniopharyngiomas.
- Childhood craniopharyngiomas are diagnosed and may be removed in the same surgery.
- Certain factors affect prognosis (chance of recovery) and treatment options.

Childhood craniopharyngiomas are benign brain tumors found near the pituitary gland.

Childhood craniopharyngiomas are rare tumors usually found near the pituitary gland (a pea-sized organ at the bottom of the brain that controls other glands) and the hypothalamus (a small cone-shaped organ connected to the pituitary gland by nerves).



Anatomy of the inside of the brain, showing the pineal and pituitary glands, optic nerve, ventricles (with cerebrospinal fluid shown in blue), and other parts of the brain.

Craniopharyngiomas are usually part solid mass and part fluid-filled cyst. They are benign (not cancer) and do not spread to other parts of the brain or to other parts of the body. However, they may grow and press on nearby parts of the brain or other areas, including the pituitary gland, the optic chiasm, optic nerves, and fluid-filled spaces in the brain. Craniopharyngiomas may affect many functions of the brain. They may affect hormone making, growth, and vision. Benign brain tumors need treatment.

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 covered in this summary. See the PDQ treatment summary on Childhood Brain and Spinal Cord Tumors Treatment Overview for information about the different types of childhood brain and spinal cord tumors.

Brain tumors can occur in both children and adults; however, treatment for children may be different than treatment for adults. (See the PDQ summary on Adult Central Nervous System Tumors Treatment for more information.)

There are no known risk factors for childhood craniopharyngioma.

Craniopharyngiomas are rare in children younger than 2 years of age and are most often diagnosed in children aged 5 to 14 years. It is not known what causes these tumors.

Signs of childhood craniopharyngioma include vision changes and slow growth.

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

- Headaches, including morning headache or headache that goes away after vomiting.
- Vision changes.
- Nausea and vomiting.
- Loss of balance or trouble walking.
- Increase in thirst or urination.
- Unusual sleepiness or change in energy level.
- Changes in personality or behavior.
- Short stature or slow growth.
- Hearing loss.
- Weight gain.

Tests that examine the brain, vision, and hormone levels are used to detect (find) childhood craniopharyngiomas.

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.
- **Visual field exam**: An exam to check a person's field of vision (the total area in which objects can be seen). This test measures both central vision (how much a person can see when looking straight ahead) and peripheral vision (how much a person can see in all other directions while staring straight ahead). Any loss of vision may be a sign of a tumor that has damaged or pressed on the parts of the brain that affect eyesight.
- **CT scan (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 x-ray 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.
- **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. A substance called gadolinium is injected into a vein. The gadolinium collects around the tumor cells so they show up brighter in the picture. This procedure is also called nuclear magnetic resonance imaging (NMRI).

- **Blood chemistry studies**: A procedure in which a blood sample is checked to measure the amounts of certain substances released into the blood by organs and tissues in the body. An unusual (higher or lower than normal) amount of a substance can be a sign of disease.
- **Blood hormone studies**: A procedure in which a blood sample is checked to measure the amounts of certain hormones released into the blood by organs and tissues in the body. An unusual (higher or lower than normal) amount of a substance can be a sign of disease in the organ or tissue that makes it. For example, the blood may be checked for unusual levels of thyroid-stimulating hormone (TSH) or adrenocorticotropic hormone (ACTH). TSH and ACTH are made by the pituitary gland in the brain.

Childhood craniopharyngiomas are diagnosed and may be removed in the same surgery.

Doctors may think a mass is a craniopharyngioma based on where it is in the brain and how it looks on a CT scan or MRI. In order to be sure, a sample of tissue is needed.

One of the following types of biopsy procedures may be used to take the sample of tissue:

- Open biopsy: A hollow needle is inserted through a hole in the skull into the brain.
- Computer-guided needle biopsy: A hollow needle guided by a computer is inserted through a small hole in the skull into the brain.
- Transsphenoidal biopsy: Instruments are inserted through the nose and sphenoid bone (a butterfly-shaped bone at the base of the skull) and into the brain.

A pathologist views the tissue under a microscope to look for tumor cells. If tumor cells are found, as much tumor as safely possible may be removed during the same surgery.

The following laboratory 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.

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

The prognosis and treatment options depend on the following:

- The size of the tumor.
- Where the tumor is in the brain.
- Whether there are tumor cells left after surgery.
- The child's age.
- Side effects that may occur months or years after treatment.
- Whether the tumor has just been diagnosed or has recurred (come back).

Stages of Childhood Craniopharyngioma

The process used to find out if cancer has spread within the brain or to other parts of the body is called staging. There is no standard system for staging childhood craniopharyngioma. Craniopharyngioma is described as newly diagnosed disease or recurrent disease.

The results of the tests and procedures done to diagnose craniopharyngioma are used to help make decisions about treatment.

Sometimes childhood craniopharyngioma recurs (comes back) after it has been treated. The tumor may come back in the same area of the brain where it was first found.

Treatment Option Overview

Key Points for This Section

- There are different types of treatment for children with craniopharyngioma.
- Children with craniopharyngioma 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.
- Treatment for childhood craniopharyngioma may cause side effects.
- Five types of treatment are used:
    - Surgery (resection)
    - Surgery and radiation therapy
    - Surgery with cyst drainage
    - Chemotherapy
    - Immunotherapy
- New types of treatment are being tested in clinical trials.
- Patients may want to think about taking part in a clinical trial.
- Patients can enter clinical trials before, during, or after starting their treatment.
- Follow-up tests may be needed.

There are different types of treatment for children with craniopharyngioma.

Different types of treatments are available for children with craniopharyngioma. 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 tumors. When clinical trials show that a new treatment is better than the standard treatment, the new treatment may become the standard treatment.

Because tumors in children are rare, taking part in a clinical trial should be considered. Clinical trials are taking place

in many parts of the country. Information about ongoing clinical trials is available from the NCI website. Choosing the most appropriate treatment is a decision that ideally involves the patient, family, and health care team.

Children with craniopharyngioma 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 tumors. The pediatric oncologist works with other pediatric healthcare 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.
- Radiation oncologist.
- Neurologist.
- Endocrinologist.
- Ophthalmologist.
- Rehabilitation specialist.
- Psychologist.
- Social worker.
- Nurse specialist.

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 diagnosis 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.

Treatment for childhood craniopharyngioma may cause side effects.

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

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

- Physical problems that affect the following:
    - Brain (seizures).
    - Bone and muscle growth and development.
- Behavior problems.
- Changes in mood, feelings, thinking, learning, or memory.
- Second cancers (new types of cancer).

The following serious physical problems may occur if the pituitary gland, hypothalamus, optic nerves, or carotid artery are affected during surgery or radiation therapy:

- Obesity.

- Metabolic syndrome, including fatty liver disease not caused by drinking alcohol.
- Vision problems, including blindness.
- Blood vessel problems or stroke.
- Loss of the ability to make certain hormones.

Some late effects may be treated or controlled. Life-long hormone replacement therapy with several medicines may be needed. It is important to talk with your child's doctors about the effects tumor treatment can have on your child. (See the PDQ summary on Late Effects of Treatment for Childhood Cancer for more information).

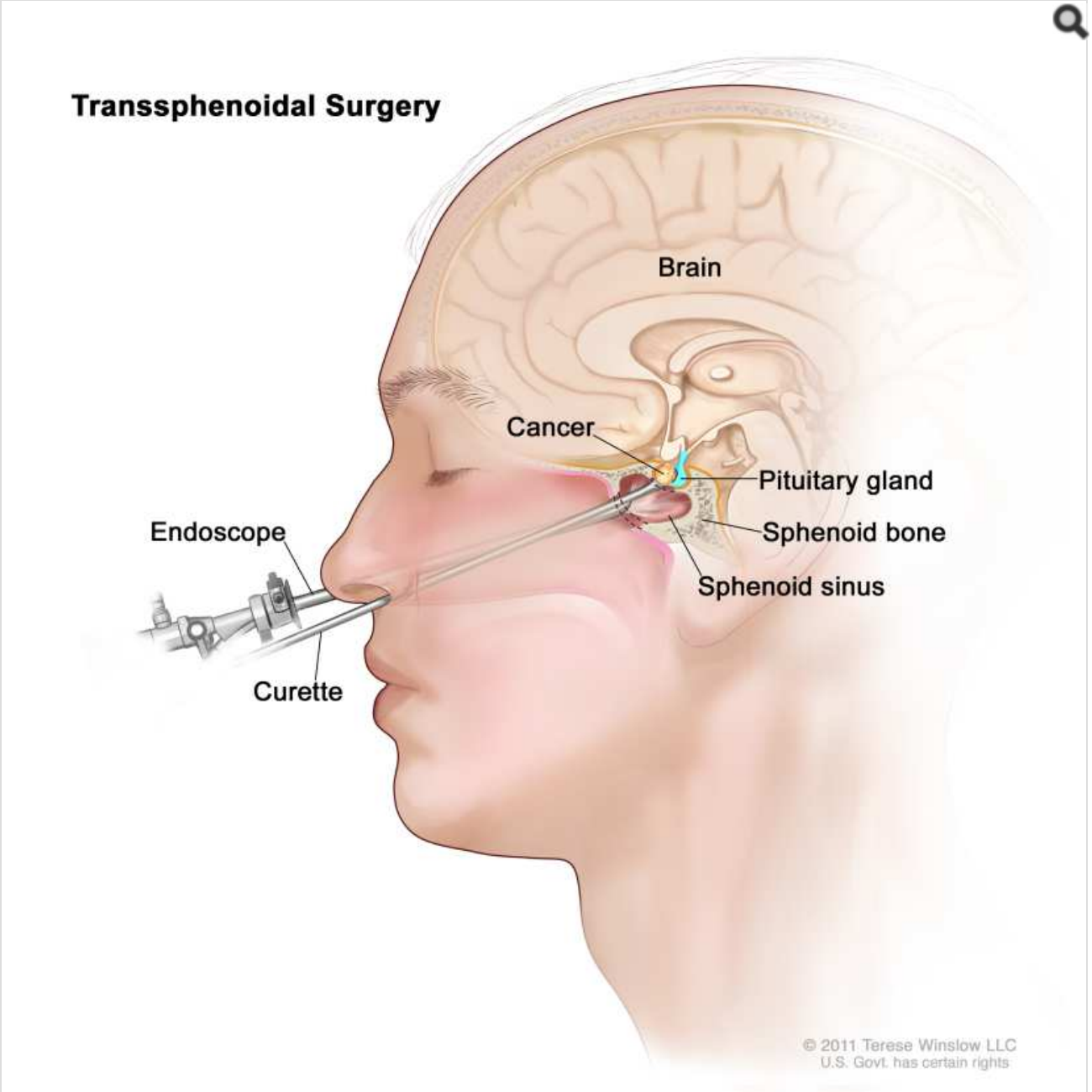
Five types of treatment are used:

Surgery (resection)

The way the surgery is done depends on the size of the tumor and where it is in the brain. It also depends on whether the tumor has grown into nearby tissue in a finger-like way and expected late effects after surgery.

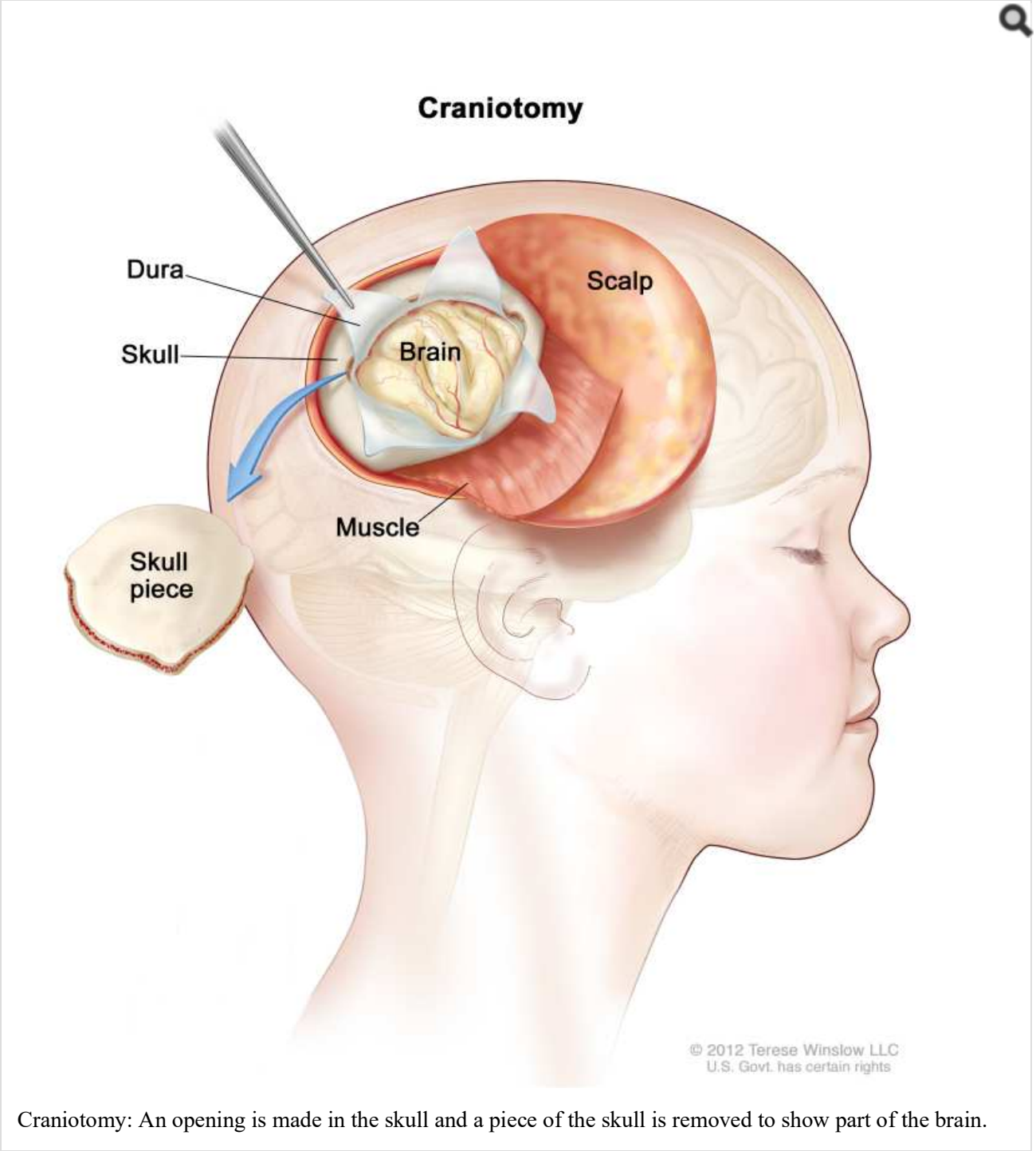
The types of surgery that may be used to remove all of the tumor that can be seen with the eye include the following:

- Transsphenoidal surgery: A type of surgery in which the instruments are inserted into part of the brain by going through an incision (cut) made under the upper lip or at the bottom of the nose between the nostrils and then through the sphenoid bone (a butterfly-shaped bone at the base of the skull) to reach the tumor near the pituitary gland and hypothalamus.



Transsphenoidal surgery. An endoscope and a curette are inserted through the nose and sphenoid sinus to remove the tumor.

- Craniotomy: Surgery to remove the tumor through an opening made in the skull.



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

Sometimes all of the tumor that can be seen is removed in surgery and no further treatment is needed. At other times, it is hard to remove the tumor because it is growing into or pressing on nearby organs. If there is tumor remaining after the surgery, radiation therapy is usually given to kill any tumor cells that are left. Treatment given after the surgery, to lower the risk that the cancer will come back, is called adjuvant therapy.

Surgery and radiation therapy

Partial resection is used to treat some craniopharyngiomas. It is used to diagnose the tumor, remove fluid from a cyst, and relieve pressure on the optic nerves. If the tumor is near the pituitary gland or hypothalamus, it is not removed. This reduces the number of serious side effects after surgery. Partial resection is followed by radiation therapy.

Radiation therapy is a tumor treatment that uses high-energy x-rays or other types of radiation to kill tumor 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 tumor.
- Internal radiation therapy uses a radioactive substance sealed in needles, seeds, wires, or catheters that are placed directly into or near the tumor.

The way the radiation therapy is given depends on the type of tumor, whether the tumor is newly diagnosed or has come back, and where the tumor formed in the brain. External and internal radiation therapy are used to treat childhood craniopharyngioma.

Because radiation therapy to the brain can affect growth and development in young children, ways of giving radiation therapy that have fewer side effects are being used. These include:

- Stereotactic radiosurgery: For very small craniopharyngiomas at the base of the brain, stereotactic radiosurgery may be used. Stereotactic radiosurgery is a type of external radiation therapy. A rigid head frame is attached to the skull to keep the head still during the radiation treatment. A machine aims a single large dose of radiation directly at the tumor. This procedure does not involve surgery. It is also called stereotaxic radiosurgery, radiosurgery, and radiation surgery.
- Intracavitary radiation therapy: Intracavitary radiation therapy is a type of internal radiation therapy that may be used in tumors that are part solid mass and part fluid-filled cyst. Radioactive material is placed inside the tumor. This type of radiation therapy causes less damage to the nearby hypothalamus and optic nerves.
- Intensity-modulated photon therapy: A type of radiation therapy that uses x-rays or gamma rays that come from a special machine called a linear accelerator (linac) to kill tumor cells. A computer is used to target the exact shape and location of the tumor. Thin beams of photons of different intensities are aimed at the tumor from many angles. This type of 3-dimensional radiation therapy may cause less damage to healthy tissue in the brain and other parts of the body. Photon therapy is different from proton therapy.
- Intensity-modulated proton therapy: A type of radiation therapy that uses streams of protons (tiny particles with a positive charge) to kill tumor cells. A computer is used to target the exact shape and location of the tumor. Thin beams of protons of different intensities are aimed at the tumor from many angles. This type of 3-dimensional radiation therapy may cause less damage to healthy tissue in the brain and other parts of the body. Proton radiation is different from x-ray radiation.

Surgery with cyst drainage

Surgery may be done to drain tumors that are mostly fluid-filled cysts. This lowers pressure in the brain and relieves symptoms. A catheter (thin tube) is inserted into the cyst and a small container is placed under the skin. The fluid drains into the container and is later removed. Sometimes, after the cyst is drained, a drug is put through the catheter into the cyst. This causes the inside wall of the cyst to scar and stops the cyst from making fluid or increases the amount of the time it takes for the fluid to build up again. Surgery to remove the tumor may be done after the cyst is drained.

Chemotherapy

Chemotherapy is a treatment that uses anticancer drugs to stop the growth of tumor 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 tumor cells throughout the body (systemic chemotherapy). When chemotherapy is placed directly into the cerebrospinal fluid or an organ, the drugs mainly affect tumor cells in those areas (regional chemotherapy).

Intracavitary chemotherapy is a type of regional chemotherapy that places drugs directly into a cavity, such as a cyst. It is used for craniopharyngioma that has come back after treatment.

Immunotherapy

Immunotherapy is a treatment that uses the patient's immune system to fight cancer. Substances made by the body or made in a laboratory are used to boost, direct, or restore the body's natural defenses against cancer. This type of cancer treatment is also called biotherapy or biologic therapy. For craniopharyngioma, the immunotherapy drug (interferon-alpha) is placed in a vein (intravenous) or inside the tumor using a catheter (intracavitary).

In newly diagnosed children, interferon-alpha may be placed directly into the cyst (intracystic) to delay the need for surgery or radiation therapy. In children whose tumor has recurred (come back), intracavitary interferon-alpha is used to treat the cyst part of the tumor.

New types of treatment are being tested in clinical trials.

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

Targeted therapy

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

Targeted therapy is being studied for the treatment of childhood craniopharyngioma that has recurred.

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 medical research process. Clinical trials are done to find out if new treatments are safe and effective or better than the standard treatment.

Many of today's standard treatments 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 diseases 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.

Use our clinical trial search 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. General information about clinical trials is also available.

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

Some clinical trials only include patients who have not yet received treatment. Other trials test treatments for patients who have not improved. There are also clinical trials that test new ways to stop a disease from recurring (coming back) or reduce the side effects of 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 disease or decide how to treat it 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 condition has changed. These tests are sometimes called follow-up tests or check-ups.

After treatment, follow-up testing with MRI will be done for several years to check if the tumor has come back.

Treatment Options for Childhood Craniopharyngioma

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

Newly Diagnosed Childhood Craniopharyngioma

Treatment of newly diagnosed childhood craniopharyngioma may include the following:

- Surgery (complete resection) with or without radiation therapy.
- Partial resection followed by radiation therapy.
- Cyst drainage with or without radiation therapy or surgery.
- Intracavitary or intracystic immunotherapy (interferon-alpha).

Use our clinical trial search 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. General information about clinical trials is also available.

Recurrent Childhood Craniopharyngioma

Craniopharyngioma may recur (come back) no matter how it was treated the first time. Treatment options for recurrent childhood craniopharyngioma depend on the type of treatment that was given when the tumor was first diagnosed and the needs of the child.

Treatment may include the following:

- Surgery (resection).
- External-beam radiation therapy.
- Stereotactic radiosurgery.
- Intracavitary radiation therapy.
- Intracavitary chemotherapy.
- Intravenous (systemic) or intracavitary immunotherapy (interferon-alpha).
- Cyst drainage.
- 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 clinical trial search 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. General information about clinical trials is also available.

To Learn More About Childhood Craniopharyngioma and Other Childhood Brain Tumors

For more information about childhood craniopharyngioma and other childhood brain 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.

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Purpose of This Summary

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Reviewers and Updates

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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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