Childhood Central Nervous System Atypical Teratoid/Rhabdoid Tumor Treatment (PDQ®) Patient Version

PDQ Pediatric Treatment Editorial Board.
Published online: September 21, 2016.
Created: December 23, 2008.

This PDQ cancer information summary has current information about the treatment of childhood central nervous system atypical teratoid and rhabdoid tumor. 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 Central Nervous System (CNS) Atypical Teratoid/Rhabdoid Tumor

Key Points for This Section

- Central nervous system atypical teratoid/rhabdoid tumor is a disease in which malignant (cancer) cells form in the tissues of the brain.
- Certain genetic changes may increase the risk of atypical teratoid/rhabdoid tumor.
- The signs and symptoms of atypical teratoid/rhabdoid tumor are not the same in every patient.
- Tests that examine the brain and spinal cord are used to detect (find) CNS atypical teratoid/rhabdoid tumor.
- Childhood atypical teratoid/rhabdoid tumor is diagnosed and may be removed in surgery.
- Certain factors affect prognosis (chance of recovery) and treatment options.

Central nervous system atypical teratoid/rhabdoid tumor is a disease in which malignant (cancer) cells form in the tissues of the brain.

Central nervous system (CNS) atypical teratoid/rhabdoid tumor (AT/RT) is a very rare, fast-growing tumor of the brain and spinal cord. It usually occurs in children aged three years and younger, although it can occur in older children and adults.

About half of these tumors form in the cerebellum or brain stem. The cerebellum is the part of the brain that controls movement, balance, and posture. The brain stem controls breathing, heart rate, and the nerves and muscles used in seeing, hearing, walking, talking, and eating. AT/RT may also be found in other parts of the central nervous system (brain and spinal cord).
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, hypothalamus, pineal gland, 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 (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).

This summary describes 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. For more information, see the PDQ summary on Childhood Brain and Spinal Cord Tumors Treatment Overview 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 treatment summary on Adult Central Nervous System Tumors Treatment for more information.

Certain genetic changes may increase the risk of atypical teratoid/rhabdoid tumor.

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

Atypical teratoid/rhabdoid tumor may be linked to a change in a tumor suppressor gene called SMARCB1. This type of gene makes a protein that helps control cell growth. Changes in the DNA of tumor suppressor genes like SMARCB1 may lead to cancer.

Changes in the SMARCB1 gene may be inherited (passed on from parents to offspring). When the SMARCB1 gene change is inherited, tumors may form in two parts of the body at the same time (for example, in the brain and the kidney). For patients with AT/RT, genetic counseling (a discussion with a trained professional about inherited diseases and a possible need for gene testing) may be recommended.

The signs and symptoms of atypical teratoid/rhabdoid tumor are not the same in every patient.

Signs and symptoms depend on the following:

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

Because atypical teratoid/rhabdoid tumor is fast growing, signs and symptoms may develop quickly and get worse over a period of days or weeks. Signs and symptoms may be caused by AT/RT or by other conditions. Check with your child's doctor if your child has any of the following:

- Morning headache or headache that goes away after vomiting.
- Nausea and vomiting.
- Unusual sleepiness or change in activity level.
- Loss of balance, lack of coordination, or trouble walking.
- Increase in head size (in infants).

**Tests that examine the brain and spinal cord are used to detect (find) CNS atypical teratoid/rhabdoid tumor.**

The following tests and procedures may be used:

- **Physical exam and 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):** 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. This procedure is also called nuclear magnetic resonance imaging (NMRI).

- **Lumbar puncture:** A procedure used to collect cerebrospinal fluid (CSF) from the spinal column. This is done by placing a needle between two bones in the spine and into the CSF around the spinal cord and removing a sample of fluid. The sample of CSF is checked under a microscope for signs of tumor cells. The sample may also be checked for the amounts of protein and glucose. A higher than normal amount of protein or lower than normal amount of glucose may be a sign of a tumor. This procedure is also called an LP or spinal tap.

- **SMARCB1 gene testing:** A laboratory test in which a sample of blood or tissue is tested for the *SMARCB1* gene.

**Childhood atypical teratoid/rhabdoid tumor is diagnosed and may be 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 of brain tumor. It is often difficult to completely remove AT/RT because of where the tumor is in the brain and because it may already have spread at the time of diagnosis.
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 test that uses antibodies to check for certain antigens in a sample of tissue. The antibody is usually linked to a radioactive substance or a dye that causes the tissue to light up under a microscope. This test is used to tell the difference between AT/RT and other brain tumors.

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

The prognosis (chance of recovery) and treatment options depend on the following:

- Whether there are certain inherited gene changes.
- The age of the child.
- The amount of tumor remaining after surgery.
Whether the cancer has spread to other parts of the central nervous system (brain and spinal cord) or to the kidney at the time of diagnosis.

**Stages of Childhood CNS Atypical Teratoid/Rhabdoid Tumor**

**Key Points for This Section**

- There is no standard staging system for central nervous system atypical teratoid/rhabdoid tumor.

**There is no standard staging system for central nervous system atypical teratoid/rhabdoid tumor.**

The extent or spread of cancer is usually described as stages. There is no standard staging system for central nervous system atypical teratoid/rhabdoid tumor.

For treatment, this tumor is grouped as newly diagnosed or recurrent. Treatment depends on the following:

- The age of the child.
- How much cancer remains after surgery to remove the tumor.

Results from the following procedure are also used to plan treatment:

- **Ultrasound exam**: A procedure in which high-energy sound waves (ultrasound) are bounced off internal tissues or organs, such as the kidney, and make echoes. The echoes form a picture of body tissues called a sonogram. The picture can be printed to be looked at later. This procedure is done to check for tumors that may also have formed in the kidney.

**Treatment Option Overview**

**Key Points for This Section**

- There are different types of treatment for patients with central nervous system atypical teratoid/rhabdoid tumor.
- Children with atypical teratoid/rhabdoid tumor should have their treatment planned by a team of health care providers who are experts in treating cancer in children.
- Childhood brain tumors may cause signs or symptoms that begin before the cancer is diagnosed and continue for months or years.
- Some cancer treatments cause side effects months or years after treatment has ended.
- Four types of treatment are used:
- 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 cancer treatment.
- Follow-up tests may be needed.

**There are different types of treatment for patients with central nervous system atypical teratoid/rhabdoid tumor.**

Different types of treatment are available for patients with central nervous system atypical teratoid/rhabdoid tumor.
Treatment for AT/RT is usually within a clinical trial. A treatment clinical trial is a research study meant to help improve current treatments or obtain information on new treatments for patients with cancer.

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 cancer treatment is a decision that ideally involves the patient, family, and health care team.

Children with atypical teratoid/rhabdoid tumor should have their treatment planned by a team of health care providers who are experts in treating cancer 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 pediatric health care providers who are experts in treating children with central nervous system cancer and who specialize in certain areas of medicine. These may include the following specialists:

- Pediatrician.
- Pediatric neurosurgeon.
- Radiation oncologist.
- Neurologist.
- Pediatric nurse specialist.
- Rehabilitation specialist.
- Psychologist.
- Social worker.
- Geneticist or genetic counselor.

**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. These signs or symptoms may 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.

**Some cancer treatments cause side effects months or years after treatment has ended.**

Side effects from cancer treatment that begin during or 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).

**Four types of treatment are used:**

**Surgery**

Surgery is used to diagnose and treat CNS atypical teratoid/rhabdoid tumor. See the General Information section of this summary.

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

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 placed directly into the cerebrospinal fluid, an organ, or a body cavity such as the abdomen, the drugs mainly affect tumor cells in those areas (regional chemotherapy). Regular doses of anticancer drugs given by mouth or vein to treat brain and spinal cord tumors cannot cross the blood-brain barrier and reach the tumor. Anticancer drugs injected into the cerebrospinal fluid are able to reach the tumor. This is called intrathecal chemotherapy.
- 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). High doses of some anticancer drugs given into a vein can cross the blood-brain barrier and reach the tumor.

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 cancer.
- Internal radiation therapy uses a radioactive substance sealed in needles, seeds, wires, or catheters that are placed directly into or near the cancer.

The way the radiation therapy is given depends on the type of tumor being treated and whether it has spread. External radiation therapy may be given to the brain and spinal cord.

Because radiation therapy can affect growth and brain development in young children, especially children who are three years old or younger, the dose of radiation therapy may be lower than in older children.

High-dose chemotherapy with stem cell transplant

High-dose chemotherapy with stem cell transplant is a method of giving high doses of chemotherapy and replacing blood-forming cells destroyed by the cancer treatment. 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 chemotherapy is completed, 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.

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 specific cancer cells. Targeted therapies usually cause less harm to normal cells than chemotherapy or radiation therapy do. Targeted therapy is being studied in the treatment of recurrent childhood central nervous system atypical teratoid/rhabdoid tumor.

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. See the Treatment Options section that follows for links to current treatment clinical trials. These have been retrieved from NCI's listing of clinical trials.

**Follow-up tests may be needed.**

Some of the tests that were done to diagnose 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 Options for Newly Diagnosed Childhood CNS Atypical Teratoid/Rhabdoid Tumor**

**Key Points for This Section**

- There is no standard treatment for patients with central nervous system atypical teratoid/rhabdoid tumor.
- Combinations of treatments are used for patients with atypical teratoid/rhabdoid tumor.

**There is no standard treatment for patients with central nervous system atypical teratoid/rhabdoid tumor.**

Because atypical teratoid/rhabdoid tumor (AT/RT) is fast-growing, a combination of treatments is usually given. After surgery to remove the tumor, treatments for AT/RT may include combinations of the following:

- Chemotherapy.
- Radiation therapy.
- High-dose chemotherapy with stem cell transplant.

Clinical trials of new treatments should be considered for patients with newly diagnosed atypical teratoid/rhabdoid tumor.

Check the list of NCI-supported cancer clinical trials that are now accepting patients with childhood atypical teratoid/rhabdoid tumor. For more specific results, refine the search by using other search features, such as the location of the trial, the type of treatment, or the name of the drug. Talk with your child's doctor about clinical trials that may be right for your child. General information about clinical trials is available from the NCI website.

**Treatment for Recurrent Childhood CNS Atypical Teratoid/Rhabdoid Tumor**

There is no standard treatment for patients with recurrent childhood central nervous system atypical teratoid/rhabdoid tumor. Treatment may include the following:
A clinical trial of targeted therapy.


Check the list of NCI-supported cancer clinical trials that are now accepting patients with childhood atypical teratoid/rhabdoid tumor. For more specific results, refine the search by using other search features, such as the location of the trial, the type of treatment, or the name of the drug. Talk with your child's doctor about clinical trials that may be right for your child. General information about clinical trials is available from the NCI website.

To Learn More about Childhood CNS Atypical Teratoid/Rhabdoid Tumor and Other Childhood Brain Tumors

For more information about childhood central nervous system atypical teratoid/rhabdoid tumor and other childhood brain tumors, see the following:

- What You Need to Know™ Brain Tumors
- Pediatric Brain Tumor Consortium (PBTC)

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

- 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 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 central nervous system atypical teratoid and rhabdoid tumor. 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 ("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.

**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 are listed in PDQ and can be found online at NCI's website. Many cancer doctors who take part in clinical trials are also listed in PDQ. For more information, call the Cancer Information Service 1-800-4-CANCER (1-800-422-6237).

**Permission to Use This Summary**

PDQ is a registered trademark. The content of PDQ documents can be used freely as text. It cannot be identified as an NCI PDQ cancer information summary unless the whole summary is shown and it is updated regularly. However, a user would be allowed to write a sentence such as “NCI’s PDQ cancer information summary about breast cancer prevention states the risks in the following way: [include excerpt from the summary].”

The best way to cite this PDQ summary is:


Images in this summary are used with permission of the author(s), artist, and/or publisher for use in the PDQ summaries only. If you want to use an image from a PDQ summary and you are not using the whole summary, you must get permission from the owner. It cannot be given by the National Cancer Institute. Information about using the images in this summary, along with many other images related to cancer can be found in Visuals Online. Visuals Online is a collection of more than 2,000 scientific images.

**Disclaimer**

The information in these summaries should not be used to make decisions about insurance reimbursement. More information on insurance coverage is available on Cancer.gov on the Managing Cancer Care page.

**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.