

National Cancer Institute

Wilms' Tumor and Other Childhood Kidney Tumors (PDQ®): Treatment

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General Information About Wilms' Tumor and Other Childhood Kidney Tumors

Wilms' tumor and other childhood kidney tumors are diseases in which malignant (cancer) cells form in the tissues of the kidney.

Wilms' tumor

Wilms' tumor and other kidney tumors are diseases in which malignant (cancer) cells are found in the kidney. In Wilms' tumor, one or more tumors may be found in one or both kidneys. There are two kidneys, one on each side of the backbone, above the waist. Tiny tubules in the kidneys filter and clean the blood, taking out waste products and making urine. The urine passes from each kidney through a long tube called a ureter into the bladder. The bladder holds the urine until it is passed from the body.

Wilms' tumor may spread to the lungs, liver, or nearby lymph nodes.

Other kidney tumors

Clear cell sarcoma of the kidney, rhabdoid tumor of the kidney, neuroepithelial tumor of the kidney, and renal cell cancer are also childhood kidney tumors, but they are not related to Wilms' tumor.

- Clear cell sarcoma of the kidney is a type of kidney tumor that may spread to the lung, bone, brain, and soft tissue.
- Rhabdoid tumor of the kidney is a type of cancer that occurs mostly in children under age 2. It grows and spreads quickly, often to the lungs and brain.
- Neuroepithelial tumors of the kidney are rare and usually occur in young adults. They grow and spread quickly.
- Renal cell cancer occurs rarely in children. It may spread to the lungs, liver, or lymph nodes.

Having certain genetic syndromes or birth defects can increase the risk of developing Wilms' tumor.

Anything that increases your risk of getting a disease is called a risk factor. Wilms' tumor may be part of a genetic syndrome that affects growth or development. A genetic syndrome is a set of symptoms or conditions that occur together and is usually caused by abnormal genes. Certain birth defects can also increase a child's risk for developing Wilms' tumor. The following genetic syndromes and birth defects have been linked to Wilms' tumor:

- WAGR (Wilms' tumor, aniridia, ambiguous genitalia, and mental retardation) syndrome.
- Beckwith-Wiedemann syndrome.
- Hemihypertrophy.
- Denys-Drash syndrome.
- Cryptorchidism.
- Hypospadias.

Children with these genetic syndromes and birth defects should be screened for Wilms' tumor every three months until age 8. An ultrasound test may be used for screening.

Possible signs of Wilms' tumor and other childhood kidney tumors include a lump in the abdomen and blood in the urine.

These and other symptoms may be caused by kidney tumors. Other conditions may cause the same symptoms. A doctor should be consulted if any of the following problems occur in the child:

- A lump, swelling, or pain in the abdomen.
- Blood in the urine.
- Fever for no known reason.

Tests that examine the kidney and the blood are used to detect (find) Wilms' tumor and other childhood kidney tumors.

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.
- Complete blood count (CBC): A procedure in which a sample of blood is drawn and checked for the following:
 - The number of red blood cells, white blood cells, and platelets.
 - The amount of hemoglobin (the protein that carries oxygen) in the red blood cells.
 - The portion of the blood sample made up of red blood cells.

- 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 in the organ or tissue that makes it.
- Liver function test: A procedure in which a blood sample is checked to measure the amounts of certain substances released into the blood by the liver. A higher than normal amount of a substance can be a sign that the liver is not working as it should.
- Renal function test: A procedure in which blood or urine samples are checked to measure the amounts of certain substances released into the blood or urine by the kidneys. A higher or lower than normal amount of a substance can be a sign that the kidneys are not working as they should.
- Urinalysis: A test to check the color of urine and its contents, such as sugar, protein, blood, and bacteria.
- Ultrasound exam: A procedure in which high-energy sound waves (ultrasound) are bounced off internal tissues or organs and make echoes. The echoes form a picture of body tissues called a sonogram. An ultrasound of the abdomen is done to diagnose a kidney tumor.
- 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.
- Abdominal x-ray: An x-ray of the organs inside the abdomen. 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.
- Biopsy: The removal of cells or tissues so they can be viewed under a microscope by a pathologist to check for signs of cancer.

Wilms' tumor and other childhood kidney tumors are usually diagnosed and removed in surgery.

Once a kidney tumor is found, surgery is done to find out whether or not the tumor is cancer. If the tumor is only in the kidney, the surgeon will remove the whole kidney (nephrectomy). If there are tumors in both kidneys or if the tumor has spread outside the kidney, a piece of the tumor will be removed. In any case, a sample of tissue from the tumor is sent to a pathologist, who looks at it under a microscope to check for signs of cancer.

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

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

- How different the tumor cells are from normal kidney cells.
- The stage of the cancer.
- The type and size of the tumor.
- The age of the child.
- Whether the tumor can be completely removed in surgery.
- Whether the cancer has just been diagnosed or has recurred (come back).
- Whether there are any abnormal chromosomes or genes.
- Whether the patient is treated by pediatric experts with experience in treating patients with Wilms' tumor.

Stages of Wilms' Tumor and Other Childhood Kidney Tumors

Wilms' tumors and other childhood kidney tumors are staged during surgery and with imaging tests.

The process used to find out if cancer has spread outside of the kidney to other parts of the body is called staging. The information gathered from the staging process determines the stage of the disease. It is important to know the stage in order to plan treatment.

For Wilms' tumors, the stage is determined during the initial surgery and with the results from imaging tests. The following imaging tests may be done to see if cancer has spread to other places in the body:

- CT scan (CAT scan): A procedure that makes a series of detailed pictures of areas inside the body, such as the chest or brain, 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.
- X-ray of the chest and bones: 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.
- 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 body, such as the brain. This procedure is also called nuclear magnetic resonance imaging (NMRI).
- Bone scan: A procedure to check if there are rapidly dividing cells, such as cancer cells, in the bone. A very small amount of radioactive material is injected into a vein and travels through the bloodstream. The radioactive material collects in the bones and is detected by a scanner.
- Ultrasound exam: A procedure in which high-energy sound waves (ultrasound) are bounced off internal tissues or organs and make echoes. The echoes form a picture of body tissues called a sonogram. An ultrasound of the major heart vessels is done to stage Wilms' tumor.

In addition to the stages, Wilms' tumors are described by their histology.

The histology (how the cells look under a microscope) of the tumor affects the prognosis and may be favorable or unfavorable. Tumors with a favorable histology respond better to treatment than those with unfavorable histology.

- Favorable histology: The cancer cells look like normal kidney cells.
- Unfavorable histology: The cancer cells are anaplastic, which means they divide rapidly and look very different from normal kidney cells. Anaplastic tumors may be focal (in one area) or diffuse (spread widely throughout an area). Focal tumors have a better prognosis than diffuse tumors.

The following stages are used for both favorable and unfavorable histology Wilms' tumors:

Stage I

In stage I, the tumor was completely removed by surgery and all of the following are true:

- Cancer was found only in the kidney and did not spread to blood vessels of the kidney.
- The outer layer of the kidney did not break open.
- The tumor did not break open.
- A biopsy of the tumor was not done.
- No cancer cells are found at the edges of the area where the tumor was removed.

Stage II

In stage II, the tumor was completely removed by surgery and no cancer cells are found at the edges of the area where the cancer was removed. Before the tumor was removed, one of the following was true:

- Cancer had spread out of the kidney to nearby soft tissue.
- Cancer had spread to blood vessels of the kidney.

Stage III

In stage III, cancer remains in the abdomen after surgery and at least one of the following is true:

- Cancer spread to lymph nodes in the abdomen or pelvis (the part of the body between the hips).
- Cancer spread to or through the surface of the peritoneum (the layer of tissue that lines the abdominal cavity and covers most organs in the abdomen).
- A biopsy of the tumor was done during surgery to remove it.

- The tumor broke open before or during surgery to remove it.
- The tumor was removed in more than one piece.

Stage IV

In stage IV, cancer has spread through the blood to organs such as the lungs, liver, bone, or brain, or to lymph nodes outside of the abdomen and pelvis.

Stage V

In stage V, cancer cells are found in both kidneys when the disease is first diagnosed. Each kidney will be staged separately as I, II, III, or IV.

Recurrent Wilms' Tumor and Other Childhood Kidney Tumors

Recurrent cancer is cancer that has recurred (come back) after it has been treated.

Treatment Option Overview

Different types of treatment are available for children with Wilms' and other childhood kidney tumors. Some treatments are standard (the currently used treatment), and some are being tested in clinical trials. A treatment clinical trial is a research study meant to help improve current treatments or obtain information on new treatments for patients with cancer. When clinical trials show that a new treatment is better than the standard treatment, the new treatment may become the standard treatment.

Because cancer in children is rare, taking part in a clinical trial should be considered. Clinical trials are taking place in many parts of the country. Information about ongoing clinical trials is available from the [NCI Web site](#)¹. Choosing the most appropriate cancer treatment is a decision that ideally involves the patient, family, and health care team.

Children with Wilms' tumor or other childhood kidney tumors should have their treatment planned by a team of doctors with expertise in treating cancer in children.

Your child's treatment will be overseen by a pediatric oncologist, a doctor who specializes in treating children with cancer. The pediatric oncologist may refer you to other doctors who have experience and expertise in treating children with Wilms' tumor or other childhood kidney tumors and who specialize in certain areas of medicine. These may include the following specialists:

- Pediatric surgeon or urologist.
- Radiation oncologist.

- Rehabilitation specialist.
- Pediatric nurse specialist.
- Social worker.

Four types of standard treatment are used:

Surgery

Wilms' tumor and other childhood kidney tumors are usually treated with nephrectomy (surgery to remove the whole kidney). Nearby lymph nodes may also be removed.

If cancer is found in both kidneys, surgery may include a partial nephrectomy (removal of the cancer in the kidney and a small amount of normal tissue around it). Partial nephrectomy is done to keep the kidney working.

Even if the doctor removes all the cancer that can be seen at the time of the surgery, some patients may be given chemotherapy or radiation therapy after surgery to kill any cancer cells that are left. Treatment given after the surgery, to increase the chances of a cure, is called adjuvant therapy. Sometimes, a second-look surgery is done to see if cancer remains after chemotherapy or radiation therapy.

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 and stage of the cancer being treated.

Chemotherapy

Chemotherapy is a cancer treatment that uses drugs to stop the growth of cancer cells, either by killing the cells or by stopping them from dividing. When chemotherapy is taken by mouth or injected into a vein or muscle, the drugs enter the bloodstream and can reach cancer cells throughout the body (systemic chemotherapy). When chemotherapy is placed directly into the spinal column, an organ, or a body cavity such as the abdomen, the drugs mainly affect cancer cells in those areas (regional chemotherapy). The way the chemotherapy is given depends on the type and stage of the cancer being treated.

Combination chemotherapy is treatment using two or more anticancer drugs.

Biologic therapy

Biologic therapy 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 immunotherapy.

New types of treatment are being tested in clinical trials. These include the following:

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 re-infused stem cells grow into (and restore) the body's blood cells.

This summary section refers to specific treatments under study in clinical trials, but it may not mention every new treatment being studied. Information about ongoing clinical trials is available from the [NCI Web site](#) ¹.

Late effects may be caused by cancer treatment.

Some cancer treatments cause side effects that continue or appear months or years after cancer treatment has ended. These are called late effects. (See the [PDQ summary on Late Effects of Treatment for Childhood Cancer](#) ² for more information.) It is important that parents of children who are treated for cancer know about the possible late effects caused by certain treatments. After several years, some patients develop another form of cancer as a result of their treatment with chemotherapy and radiation. Clinical trials are ongoing to find out if lower doses of chemotherapy and radiation can be used.

Treatment Options for Wilms' Tumor and Other Childhood Kidney Tumors

Stage I Wilms' Tumor

Treatment of stage I Wilms' tumor with either favorable or unfavorable histology may include the following:

- Nephrectomy with lymph node removal, followed by combination chemotherapy.

- A clinical trial of nephrectomy alone.

This summary section refers to specific treatments under study in clinical trials, but it may not mention every new treatment being studied. Information about ongoing clinical trials is available from the [NCI Web site](#)¹.

Stage II Wilms' Tumor

Treatment of stage II Wilms' tumor with favorable histology is usually nephrectomy with removal of lymph nodes, followed by combination chemotherapy.

Treatment of stage II Wilms' tumor with unfavorable histology is usually nephrectomy with removal of lymph nodes, followed by radiation therapy to the abdomen and combination chemotherapy.

Stage III Wilms' Tumor

Treatment of stage III Wilms' tumor with either favorable or unfavorable histology is usually nephrectomy with removal of lymph nodes, followed by radiation therapy to the abdomen and combination chemotherapy.

Stage IV Wilms' Tumor

Treatment of stage IV Wilms' tumor with either favorable or unfavorable histology is usually nephrectomy with removal of lymph nodes, followed by radiation therapy to the abdomen and combination chemotherapy. Some patients may also receive radiation therapy to the lungs.

Stage V Wilms' Tumor

Treatment of stage V Wilms' tumor may be different for each patient. Treatment is usually chemotherapy to shrink the tumor, followed by surgery to remove as much of the cancer as possible. This may be followed by more chemotherapy and/or radiation therapy if cancer remains after surgery.

Inoperable Tumors

Sometimes the tumor is inoperable (cannot be removed by surgery) because it is too close to important organs or blood vessels or because it is too large to remove. In this case, chemotherapy may be given to reduce the size of the tumor so it may be removed in surgery. If the tumor does not shrink enough after chemotherapy, radiation therapy may be given to shrink it further so that surgery may be done. This may be followed by more chemotherapy and/or more radiation therapy.

Clear Cell Sarcoma of the Kidney

There is no standard treatment for clear cell sarcoma of the kidney. Treatment is usually within a clinical trial and may include nephrectomy, followed by radiation therapy to the abdomen and combination chemotherapy. Some patients may also receive radiation therapy to the lungs.

This summary section refers to specific treatments under study in clinical trials, but it may not mention every new treatment being studied. Information about ongoing clinical trials is available from the [NCI Web site](#) ¹.

Rhabdoid Tumor of the Kidney

There is no standard treatment for rhabdoid tumor of the kidney. Treatment may be within a clinical trial and may include combination chemotherapy.

This summary section refers to specific treatments under study in clinical trials, but it may not mention every new treatment being studied. Information about ongoing clinical trials is available from the [NCI Web site](#) ¹.

Neuroepithelial Tumor of the Kidney

There is no standard treatment for neuroepithelial tumor of the kidney. Treatment is usually within a clinical trial. It may be treated in the same way that Ewing's family of tumors or primitive neuroectodermal tumors are treated. See the PDQ summary on [Ewing's Family of Tumors Treatment](#) ³ for more information.

Information about ongoing clinical trials is available from the [NCI Web site](#) ¹.

Renal Cell Cancer

Treatment of renal cell cancer is usually nephrectomy with removal of lymph nodes. If cancer has spread, treatment may include biologic therapy or surgery to remove the primary tumor. See the PDQ summary on [Renal Cell Cancer Treatment](#) ⁴ for more information.

Recurrent Wilms' Tumor and Other Childhood Kidney Tumors

Treatment of recurrent Wilms' tumor may be within a clinical trial of combination chemotherapy, surgery, and radiation therapy, with or without stem cell transplant, using the child's own blood stem cells.

Treatment of recurrent clear cell sarcoma, rhabdoid tumor, and neuroepithelial tumor of the kidney is usually within a clinical trial.

This summary section refers to specific treatments under study in clinical trials, but it may not mention every new treatment being studied. Information about ongoing clinical trials is available from the [NCI Web site](#) ¹.

Changes to This Summary (11/27/2006)

The [PDQ cancer](#) information summaries are reviewed regularly and updated as new information becomes available. This section describes the latest changes made to this summary as of the date above.

Editorial changes were made to this summary.

To Learn More

Call

For more information, U.S. residents may call the National Cancer Institute's (NCI's) Cancer Information Service toll-free at 1-800-4-CANCER (1-800-422-6237) Monday through Friday from 9:00 a.m. to 4:30 p.m. Deaf and hard-of-hearing callers with TTY equipment may call 1-800-332-8615. The call is free and a trained Cancer Information Specialist is available to answer your questions.

Web sites and Organizations

The [NCI Web site](#) ⁵ provides online access to information on cancer, clinical trials, and other Web sites and organizations that offer support and resources for cancer patients and their families. There are also many other places where people can get materials and information about cancer treatment and services. Local hospitals may have information on local and regional agencies that offer information about finances, getting to and from treatment, receiving care at home, and dealing with problems associated with cancer treatment.

Publications

The NCI has booklets and other materials for patients, health professionals, and the public. These publications discuss types of cancer, methods of cancer treatment, coping with cancer, and clinical trials. Some publications provide information on tests for cancer, cancer causes and prevention, cancer statistics, and NCI research activities. NCI materials on these and other topics may be ordered online or printed directly from the [NCI Publications Locator](#) ⁶. These materials can also be ordered by telephone from the Cancer Information Service toll-free at 1-800-4-CANCER (1-800-422-6237), TTY at 1-800-332-8615.

LiveHelp

The NCI's LiveHelp service, a program available on several of the Institute's Web sites, provides Internet users with the ability to chat online with an Information Specialist. The service is available from 9:00 a.m. to 11:00 p.m. Eastern time, Monday through Friday. Information Specialists can help Internet users find information on NCI Web sites and answer questions about cancer.

Write

For more information from the NCI, please write to this address:

NCI Public Inquiries Office
Suite 3036A
6116 Executive Boulevard, MSC8322
Bethesda, MD 20892-8322

About PDQ

PDQ is a comprehensive cancer database available on NCI's Web site.

PDQ is the National Cancer Institute's (NCI's) comprehensive cancer information database. Most of the information contained in PDQ is available online at [NCI's Web site](#)⁵. PDQ is provided as a service of the NCI. The NCI is part of the National Institutes of Health, the federal government's focal point for biomedical research.

PDQ contains cancer information summaries.

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 are available in two versions. The health professional versions provide detailed information written in technical language. The patient versions are written in easy-to-understand, nontechnical language. Both versions provide current and accurate cancer information.

The PDQ cancer information summaries are developed by cancer experts and reviewed regularly.

Editorial Boards made up of experts in oncology and related specialties are responsible for writing and maintaining the cancer information summaries. The summaries are reviewed regularly and changes are made as new information becomes available. The date on each summary ("Date Last Modified") indicates the time of the most recent change.

PDQ also contains information on clinical trials.

In the United States, about two-thirds of children with cancer are treated in a clinical trial at some point in their illness. 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 new treatments, the risks involved, and how well they do or do not work. If a clinical trial shows that a new treatment is better than one currently being used, the new treatment may become "standard."

Listings of clinical trials are included in PDQ and are available online at [NCI's Web site](#)⁷. Descriptions of the trials are available in health professional and patient versions. For additional help in locating a childhood cancer clinical trial, call the Cancer Information Service at 1-800-4-CANCER (1-800-422-6237), TTY at 1-800-332-8615.

The PDQ database contains listings of groups specializing in clinical trials.

The Children's Oncology Group (COG) is the major group that organizes clinical trials for childhood cancers in the United States. Information about contacting COG is available on the [NCI Web site](#)⁵ or from the Cancer Information Service at 1-800-4-CANCER (1-800-422-6237), TTY at 1-800-332-8615.

The PDQ database contains listings of cancer health professionals and hospitals with cancer programs.

Because cancer in children and adolescents is rare, the majority of children with cancer are treated by health professionals specializing in childhood cancers, at hospitals or cancer centers with special facilities to treat them. The PDQ database contains listings of health professionals who specialize in childhood cancer and listings of hospitals with cancer programs. For help locating childhood cancer health professionals or a hospital with cancer programs, call the Cancer Information Service at 1-800-4-CANCER (1-800-422-6237), TTY at 1-800-332-8615.

<http://www.cancer.gov/cancertopics/pdq/treatment/wilms/patient>

<http://www.cancer.gov/cancertopics/pdq/treatment/wilms/patient/allpages/print>

Table of Links

¹ <http://cancer.gov/clinicaltrials>

² <http://cancer.gov/cancertopics/pdq/treatment/lateeffects/Patient>

³ <http://cancer.gov/cancertopics/pdq/treatment/ewings/Patient>

⁴ <http://cancer.gov/cancertopics/pdq/treatment/renalcell/Patient>

⁵ <http://cancer.gov>

⁶ <https://cissecure.nci.nih.gov/ncipubs>

⁷ http://cancer.gov/clinical_trials