About Wilms Tumor

Overview and Types

If your child has been diagnosed with a Wilms tumor or you are worried about it, you likely have a lot of questions. Learning some basics is a good place to start.

- What Are Wilms Tumors?

Research and Statistics

See the latest estimates for new cases of Wilms tumor in the US and what research is currently being done.

- Key Statistics for Wilms Tumors
- What’s New in Wilms Tumor Research?

What Are Wilms Tumors?

Wilms tumor (also called Wilms’ tumor or nephroblastoma) is a type of childhood cancer that starts in the kidneys. It is the most common type of kidney cancer in children. About 9 of 10 kidney cancers in children are Wilms tumors.

Cancer starts when cells in the body begin to grow out of control. Cells in nearly any part of the body can become cancer, and can spread to other areas of the body. To learn more about how cancers start and spread, see What Is Cancer? For information
about the differences between childhood cancers and adult cancers, see Cancer in Children\(^2\).

## About the kidneys

The kidneys are 2 bean-shaped organs that are attached to the back wall of the abdomen, just to the left and right of the backbone. The kidneys do a number of things:

- They filter the blood to remove excess water, salt, and waste products, which leave the body as urine.
- They help control blood pressure.
- They help make sure the body has enough red blood cells.

On top of each kidney is a small gland called an adrenal gland. The adrenal glands make hormones that have many functions, including helping the body burn fat and protein and respond to stress.

Each kidney and adrenal gland is surrounded by fat and a thin, fibrous capsule (known as Gerota’s fascia). They are protected by the lower rib cage.

Our kidneys are important, but we actually need less than one complete kidney to do all of its basic functions. Many people in the United States live normal, healthy lives with just one kidney.
Most Wilms tumors are **unilateral**, which means they affect only one kidney. Most often there is only one tumor, but a small number of children with Wilms tumors have more than one tumor in the same kidney. About 5% to 10% of children with Wilms tumors have **bilateral** disease (tumors in both kidneys).

Wilms tumors often become quite large before they are noticed. The average newly found Wilms tumor is many times larger than the kidney in which it started. Most Wilms tumors are found before they have spread (metastasized) to other organs.

**Types of Wilms tumor**

Wilms tumors are grouped into 2 major types based on how they look under a microscope (their histology):

- **Favorable histology:** The cancer cells in these tumors don’t look quite normal, but there is no anaplasia (see next paragraph). About 9 of 10 Wilms tumors have a favorable histology. The chance of curing children with these tumors is very good.
- **Anaplastic histology:** In these tumors, the look of the cancer cells varies widely, and the cells’ nuclei (the central parts that contain the DNA) tend to be very large and distorted. This is called **anaplasia**. In general, tumors in which the anaplasia is spread throughout the tumor (known as **diffuse anaplasia**) are harder to treat than tumors in which the anaplasia is limited just to certain parts of the tumor (known as **focal anaplasia**).

**Other types of kidney cancers in children**

In rare cases, children can develop other types of kidney tumors.

**Mesoblastic nephroma**

These tumors usually appear in the first few months of life. Children are usually cured with **surgery**, but sometimes **chemotherapy** is given as well. These tumors sometimes come back soon after treatment, so children who have had these tumors need to be watched closely for the first year afterward.

**Clear cell sarcoma of kidney (CCSK)**

These tumors are much more likely to spread to other parts of the body than Wilms...
tumors, and they are harder to cure. Because these tumors are rare, treatment is often given as part of a clinical trial. It's usually much like the intensive treatment used for Wilms tumors with anaplastic histology (see Treatment of Wilms Tumors by Type and Stage).

**Malignant rhabdoid tumor of the kidney**

These tumors occur most often in infants and toddlers. They tend to spread to other parts of the body quickly, and most have already spread by the time they are found, which makes them hard to cure. Because these tumors are rare, treatment is often given as part of a clinical trial, and usually includes chemotherapy with several different drugs.

**Renal cell carcinoma**

This is the most common type of kidney cancer in adults, but it also accounts for a small number of kidney cancers in children. It’s rare in young children, but it’s actually more common than Wilms tumor in older teens.

Surgery to remove the kidney (or just the tumor) is the main treatment for these cancers if it can be done. The outlook for these cancers depends largely on the extent (stage) of the cancer at the time it’s found, whether it can be removed completely with surgery, and its subtype (based on how the cancer cells look under a microscope). If the cancer is too advanced to be removed by surgery, other types of treatment may be needed.

**Hyperlinks**


References


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Key Statistics for Wilms Tumors

Wilms tumor is the most common type of kidney cancer in children. Each year, about 500 to 600 new cases of Wilms tumor are diagnosed in the United States. This number has been fairly stable for many years. About 5% of all cancers in children are Wilms tumors.

Wilms tumors tend to occur in young children. The average age at diagnosis is about 3 to 4 years. These tumors become less common as children grow older. They're very rare in adults, although cases have been reported.

Wilms tumors are slightly more common in girls than in boys. The risk of Wilms tumor is slightly higher in African-American children than in white children and is lowest among Asian-American children.

Statistics related to survival for Wilms tumors are discussed in Survival Rates for Wilms
Tumors\(^1\).

Visit the American Cancer Society’s Cancer Statistics Center\(^2\) for more key statistics.

Hyperlinks

2. [https://cancerstatisticscenter.cancer.org](https://cancerstatisticscenter.cancer.org)

References


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What’s New in Wilms Tumor Research?

Over the past few decades, research into Wilms tumor has led to many advances and much higher cure rates for this type of cancer. Still, not all children are cured, and even those who are cured might still have long-term side effects\(^1\) from treatment, so more research is needed.
In the United States, much of the research on Wilms tumor is coordinated by the Children’s Oncology Group (COG), whose main goal is to improve the treatment and quality of life of children with Wilms tumor and other types of cancer. COG is a large group of doctors, nurses, scientists, and other health professionals whose hard work has already saved the lives of many children with Wilms tumors.

**Genetics of Wilms tumors**

Research is continuing to unravel how changes in certain genes cause Wilms tumors and affect how aggressive these tumors are likely to be.

As doctors have learned how to treat Wilms tumors more effectively, they have begun to look for ways to determine which children might need more aggressive treatment to be cured, and which children might be spared from more intense treatment (and its possible side effects). For example, studies have shown that Wilms tumors with certain changes on chromosomes 1 or 16 seem to be more likely to come back after treatment. Doctors are now studying whether children with such tumors might benefit from more intense treatment.

Researchers are also studying the gene changes that seem to cause Wilms tumor cells to grow and spread. This may lead to treatments that specifically target these changes.

**Treatment of Wilms tumors**

Researchers continue to study ways to improve treatment for children with Wilms tumors.

**Using less treatment when possible**

Earlier studies found treatments that were very effective in curing most Wilms tumors, especially those with favorable histology. Current clinical trials are studying ways to treat these cancers successfully while reducing side effects as much as possible. For example:

- Studies are looking at whether young children who have Wilms tumors with very favorable features need any treatment other than surgery.
- Recent research has suggested that some children with tumors that have spread to only one spot in the lung might not need radiation therapy to the lungs, which can cause long-term side effects.
- Other research is looking at whether some tumors in the lung can be treated with
stereotactic body radiation therapy (SBRT). In this type of radiation therapy, a large dose of radiation is focused very tightly on the tumor(s), as opposed to treating the whole lung.

Newer approaches for tumors that are harder to treat

The outlook for some children with Wilms tumors, such as those with anaplastic histology, is not as good, and doctors are looking for better treatments for these children.

Newer chemotherapy drugs such as topotecan and irinotecan are now being tested.

Other studies are looking at stem cell transplants (also known as bone marrow transplants), which let doctors give higher doses of chemo than the body normally could tolerate. This approach might help treat tumors that are not responding to standard treatments or that would otherwise have a poor outlook.

As researchers have learned more about the gene changes in cancer cells, they have started to develop newer drugs that specifically target these changes. Targeted drugs work differently from standard chemotherapy drugs. They sometimes work when chemo drugs don’t, and they often have different (and less severe) side effects. Targeted therapies have already become standard treatments for some kinds of cancers.

For children with Wilms tumors who might not be cured with current treatments, some clinical trials are now testing the tumor cells for certain gene changes. If one of these changes is found, treating the tumor with a drug that can target the change might be helpful. About a dozen different targeted drugs are now being tested in this way.

Hyperlinks

2. [https://childrensoncologygroup.org/](https://childrensoncologygroup.org/)

References


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us/policies/content-usage.html).
Wilms Tumor Causes, Risk Factors, and Prevention

Risk Factors

A risk factor is anything that affects your chance of getting a disease such as cancer. Learn more about the risk factors for Wilms tumors.

- Risk Factors for Wilms Tumors
- What Causes Wilms Tumors?

Prevention

There are no known lifestyle-related or environmental causes of Wilms tumors, so at this time there is no way to protect against most of these cancers.

- Can Wilms Tumors Be Prevented?

Risk Factors for Wilms Tumors

A risk factor is anything that raises a person’s chance of having a disease such as cancer. Different cancers have different risk factors.

Lifestyle-related risk factors such as body weight, physical activity, diet, and tobacco use play a major role in many adult cancers. But these factors usually take many years
to influence cancer risk, and they are not thought to have much of an effect on the risk of childhood cancers, including Wilms tumors.

So far, research hasn't found any strong links between Wilms tumor and environmental factors, either during a mother's pregnancy or after a child's birth.

Most Wilms tumors have no clear cause, but there are some factors that affect risk.

**Age**

Wilms tumors are most common in young children, with the average age being about 3 to 4 years. They are less common in older children, and rare in adults.

**Race/ethnicity**

In the United States, the risk of Wilms tumor is slightly higher in African-American children than in white children and is lowest among Asian-American children. The reason for this is not known.

**Gender**

Wilms tumors are slightly more common in girls than in boys.

**Family history of Wilms tumor**

About 1% to 2% of children with Wilms tumors have one or more relatives with the same cancer. Scientists think that these children inherit chromosomes with an abnormal or missing gene\(^1\) from a parent that increases their risk of developing Wilms tumor.

Children with a family history of Wilms tumors are slightly more likely to have tumors in both kidneys. Still, in most children only one kidney is affected.

**Certain genetic syndromes/birth defects**

There is a strong link between Wilms tumors and certain kinds of birth defects. About 1 child in 10 with Wilms tumor also has birth defects. Most birth defects linked to Wilms tumors occur in syndromes. A syndrome is a group of symptoms, signs, malformations, or other abnormalities that occur together in the same person. Syndromes linked to Wilms tumor include:
WAGR syndrome

WAGR stands for the first letters of the physical and mental problems linked with this syndrome (although not all children have all of them):

- Wilms tumor
- Aniridia (complete or partial lack of the iris [colored area] of the eyes)
- Genitourinary tract abnormalities (defects of the kidneys, urinary tract, penis, scrotum, clitoris, testicles, or ovaries)
- Mental Retardation

Children with this syndrome have about a 30% to 50% chance of having a Wilms tumor. The cells in children with WAGR syndrome are missing part of chromosome 11, where the \( WT1 \) gene is normally found (see What Causes Wilms Tumors?). Children with WAGR tend to get Wilms tumors at an earlier age and often have tumors in both kidneys.

Denys-Drash syndrome and Frasier syndrome

These rare syndromes have also been linked to changes (mutations) in the \( WT1 \) gene.

In Denys-Drash syndrome, the kidneys become diseased and stop working when the child is very young. Wilms tumors usually develop in the diseased kidneys. The reproductive organs don’t develop normally, and boys may be mistaken for girls. Because the risk of Wilms tumors is very high, doctors often advise removing the kidneys soon after this syndrome is diagnosed.

In Frasier syndrome the kidneys are also diseased, but they usually keep working into adolescence. As with Denys-Drash syndrome, the reproductive organs don’t develop normally. Children with Frasier syndrome are also at increased risk for Wilms tumors, although they are at even higher risk for cancers in the reproductive organs.

Beckwith-Wiedemann syndrome

Children with this syndrome tend to be big for their age. They also have larger than normal internal organs and often have an enlarged tongue. They may have an oversized arm and/or leg on one side of the body (called hemihypertrophy), as well as other medical problems. They have about a 5% risk of having Wilms tumors (or, less often, other cancers that develop during childhood). This syndrome is caused by a defect in chromosome 11 that affects the \( WT2 \) gene.
Other syndromes

Less often, Wilms tumor has been linked to other syndromes, including:

- Perlman syndrome
- Sotos syndrome
- Simpson-Golabi-Behmel syndrome
- Bloom syndrome
- Li-Fraumeni syndrome
- Trisomy 18

Certain birth defects

Wilms tumor is also more common in children with certain birth defects (without known syndromes):

- Aniridia (complete or partial lack of the iris [colored area] of the eyes)
- Hemihypertrophy (an oversized arm and/or leg on one side of the body)
- Cryptorchidism (failure of the testicles to descend into the scrotum) in boys
- Hypospadias (defect in boys where the urinary opening is on the underside of the penis)

Hyperlinks


References


National Cancer Institute. Wilms Tumor and Other Childhood Kidney Tumors Treatment
What Causes Wilms Tumors?

Although there is a clear link between Wilms tumors and certain birth defect syndromes and genetic changes, most children with this type of cancer do not have any known birth defects or inherited gene changes.

Researchers do not yet know exactly why some children get Wilms tumors, but they have made great progress in understanding how normal kidneys develop, as well as how this process can go wrong, leading to a Wilms tumor.

The kidneys develop very early as a fetus grows in the womb. Some of the cells that are supposed to develop into mature kidney cells sometimes stay as early kidney cells instead, and might remain even after the baby is born. Usually, these cells mature by the time the child is 3 to 4 years old. But if this doesn’t happen, the cells might somehow begin to grow out of control, which might result in a Wilms tumor.

Changes in genes

Normal human cells grow and function based mainly on the information contained in each cell’s DNA. DNA is the chemical in each of our cells that makes up our genes, which control how our cells function. Genes are packaged in chromosomes (long strands of DNA). We normally have 23 pairs of chromosomes in each cell (with one set of chromosomes coming from each parent). We usually look like our parents because they are the source of our DNA. But DNA affects more than how we look.

Some genes control when our cells grow, divide into new cells, and die:

- Genes that help cells grow, divide, or stay alive are called oncogenes.
- Genes that slow down cell division or cause cells to die at the right time are called tumor suppressor genes.
Cancers can be caused by DNA changes (mutations) that turn on oncogenes or turn off tumor suppressor genes. Changes in certain genes in early kidney cells can lead to problems as the kidneys develop.

Sometimes these gene changes are passed on from a parent to a child, but most Wilms tumors don’t seem to be caused by inherited gene mutations. Instead, they seem to result from gene changes that occur early in a child’s life, perhaps even before birth.

**Gene changes in Wilms tumors**

Doctors have found that some Wilms tumors have changes in specific genes:

- A small number of Wilms tumors have changes in or loss of the *WT1* or *WT2* genes, which are tumor suppressor genes found on chromosome 11. Changes in these genes and some other genes on chromosome 11 can lead to overgrowth of certain body tissues. This may explain why some other growth abnormalities, like those described in Risk Factors for Wilms Tumors, are sometimes found along with Wilms tumors.
- In a small number of Wilms tumors there is a change in a tumor suppressor gene known as *WTX*, which is found on the X chromosome.
- Another gene that is sometimes altered in Wilms tumor cells is known as *CTNNB1*, which is on chromosome 3.

It’s not clear exactly what causes these genes to be altered.

Several other gene or chromosome changes have been found in Wilms tumor cells. Typically, more than one gene change is needed to cause cancer. None of the gene changes found so far are seen in all Wilms tumors. There are also likely to be other gene changes that have not yet been found.

Researchers now understand some of the gene changes that can occur in Wilms tumors, but it’s still not clear what causes these changes. Some gene changes can be inherited, but most Wilms tumors are not the result of known inherited syndromes.

Some gene changes may just be random events that sometimes happen inside a cell, without having an outside cause. There are no known lifestyle-related or environmental causes of Wilms tumors, so it’s important to know that there is nothing these children or their parents could have done to prevent these cancers.
Can Wilms Tumors Be Prevented?

The risk of many adult cancers can be reduced with certain lifestyle changes (such as staying at a healthy weight or quitting smoking), but at this time there are no known ways to prevent most cancers in children.

The only known risk factors for Wilms tumors (age, race, gender, and certain inherited conditions) can’t be changed. There are no known lifestyle-related or environmental causes of Wilms tumors, so at this time there is no way to prevent most of these cancers. Experts think these cancers come from cells that were around before birth but failed to develop into mature kidney cells. This doesn’t seem to be caused by anything a mother could avoid during pregnancy.

For children at very high risk

In some very rare cases, such as in children with Denys-Drash syndrome who are almost certain to develop Wilms tumors, doctors may recommend removing the kidneys at a very young age (with a donor kidney transplant later on) to prevent tumors from developing.
References


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Wilms Tumor Early Detection, Diagnosis, and Staging

Detection and Diagnosis

Catching cancer early often allows for more treatment options. Some early cancers may have signs and symptoms that can be noticed, but that is not always the case.

- Can Wilms Tumors Be Found Early?
- Signs and Symptoms of Wilms Tumors
- Tests for Wilms Tumors

Stages and Outlook (Prognosis)

After a cancer diagnosis, staging provides important information about the extent of cancer in the body and anticipated response to treatment.

- Wilms Tumor Stages
- Survival Rates for Wilms Tumors

Questions to Ask About Wilms Tumor

Here are some questions you can ask your cancer care team to help you better understand your cancer diagnosis and treatment options.

- Questions to Ask Your Child’s Doctor About Wilms Tumor
Can Wilms Tumors Be Found Early?

Wilms tumors are usually found when they start to cause symptoms such as swelling in the abdomen (belly), but by this point they have often grown quite large. They can be found earlier in some children with tests such as an ultrasound of the abdomen. (See Tests for Wilms Tumors.) But because Wilms tumors are rare, it’s not practical to use ultrasound exams to screen all children for them. (Screening is testing for a disease like cancer in people with no signs or symptoms.) There are no blood tests or other tests that are useful in screening otherwise healthy children for Wilms tumors.

For children at increased risk

On the other hand, screening is very important for children who have syndromes or birth defects known to be linked to Wilms tumors. For these children, most doctors recommend physical exams by a specialist and ultrasound exams of the kidneys on a regular basis (for example, about every 3 or 4 months at least until the age of 7) to find any kidney tumors when they are still small and have not yet spread to other organs.

Wilms tumor can also run in families, although this is rare. Talk to your doctor if you have any relatives who have had a Wilms tumor. If you do, the children in your family may need to have regular ultrasound exams of the abdomen. If a man or woman is known to have a WT1 gene mutation, genetic testing can be done to see if they have passed the mutation on to their children. (This can be done even before birth.)

Hyperlinks

2. www.cancer.org/cancer/cancer-causes/genetics.html

References


Signs and Symptoms of Wilms Tumors

Wilms tumors occur most often in young children. These tumors often grow quite large before causing any symptoms. Children may look healthy and act and play normally.

Swelling or a large lump in the abdomen (belly)

This is often the first sign of a Wilms tumor. Parents may notice swelling or hardness in the belly while bathing or dressing the child. The lump is sometimes large enough to be felt on both sides of the belly. It’s usually not painful, but it might be in some children.

Other possible symptoms

Some children with Wilms tumor may also have:

- Fever
- Nausea
- Loss of appetite
- Shortness of breath
- Constipation
- Blood in the urine

Wilms tumors can also sometimes cause high blood pressure. This doesn’t usually cause symptoms on its own, but in rare cases blood pressure can get high enough to cause problems such as headaches, bleeding inside the eye, or even a change in consciousness.

Many of the signs and symptoms of Wilms tumors are more likely to be caused by something else. Still, if your child has any of these symptoms, check with your child’s doctor so that the cause can be found and treated, if needed.
Tests for Wilms Tumors

Certain signs and symptoms could suggest that a child might have a Wilms tumor (or another type of kidney tumor), but exams and tests are needed to find out for sure.

Medical history and physical exam

If your child has signs or symptoms that suggest he or she might have a kidney tumor, the doctor will want to get a complete medical history to learn more about the symptoms and how long they have been there. The doctor may also ask if there’s a family history of cancer or birth defects, especially in the genitals or urinary system, as this might point to an increased risk or Wilms tumors.

The doctor will also do a physical exam to look for possible signs of a kidney tumor or other health problems. The focus will probably be on your child’s abdomen (belly) and on any increase in blood pressure, which is another possible sign of a kidney tumor. Blood and urine samples might also be collected and tested (see “Lab tests” below).

Imaging tests
If the doctor thinks your child might have a kidney tumor, he or she will probably get one or more imaging tests. These tests use sound waves, x-rays, magnetic fields, or radioactive substances to create pictures of the inside of the body. Imaging tests might be done for a number of reasons, including:

- To help find out if there is a tumor in the kidney(s), and if so, if it's likely to be a Wilms tumor
- To learn if and how far the tumor has spread, both in the kidney(s) and to other parts of the body
- To help guide surgery or radiation therapy
- To look at the area after treatment to help determine if it has worked

**Ultrasound (sonogram)**

Ultrasound\(^2\) is often the first imaging test done if the doctor suspects your child has a tumor in the abdomen. This test is easy to have, does not use radiation, and it gives the doctor a good view of the kidneys and the other organs in the abdomen.

This test can also show if the tumor is growing into the main veins coming out of the kidney. This can help in planning for surgery, if it's needed.

**Computed tomography (CT, CAT) scan**

The CT scan\(^3\) uses x-rays to make detailed cross-sectional images of parts of your child’s body, including the kidneys. This is one of the most useful tests to look for a tumor inside the kidney. It’s also helpful for checking whether a cancer has grown into nearby veins or has spread to organs beyond the kidney, such as the lungs. Your child will need to lie very still on a table while the scans are being done. Younger children may be given medicine to help keep them calm or even asleep during the test to help make sure the pictures are clear.

**Magnetic resonance imaging (MRI) scan**

An MRI scan\(^4\) creates detailed images using radio waves and strong magnets instead of x-rays, so there is no radiation involved. This test might be done if the doctor needs to see very detailed images of the kidney or nearby areas. For example, it might be done if there’s a chance that a kidney tumor might have reached a major vein (the inferior vena cava) in the abdomen. An MRI might also be used to look for possible spread of cancer to the brain or spinal cord if doctors are concerned the cancer may have spread there.
Your child may have to lie inside a narrow tube, which is confining and can be distressing. The test also requires a person to stay still for several minutes at a time. Younger children may be given medicine to help keep them calm or even asleep during the test.

**Chest x-ray**

Chest x-rays\(^5\) may be done to look for any spread of Wilms tumor to the lungs, as well as to have a baseline view of the lungs to compare with other x-rays that might be done in the future. This test might not be needed if a CT scan of the chest is done.

**Lab tests**

Lab tests\(^6\) might be done to check urine and blood samples if your child’s doctor suspects a kidney problem. They may also be done after a Wilms tumor has been found.

A urine sample may be tested (urinalysis) to see if there are problems with the kidneys. Urine may also be tested for substances called catecholamines. This is done to make sure your child doesn’t have another kind of tumor called neuroblastoma\(^7\). (Neuroblastomas often start in the adrenal glands, which are just on top of each kidney.)

**Kidney biopsy/surgery**

Most of the time, imaging tests can give doctors enough information to decide if a child probably has a Wilms tumor, and therefore if surgery\(^8\) should be done. But the actual diagnosis of Wilms tumor is made when a small piece of the tumor is removed and checked under a microscope. The cells in Wilms tumors have a distinct appearance when looked at this way. Doctors also look at the sample to determine the histology of the Wilms tumor (favorable or anaplastic), as described in What Are Wilms Tumors?\(^9\)

Most often, a sample is removed during surgery\(^10\) to treat the tumor. Sometimes if the doctors are less certain about the diagnosis or if they aren’t sure the tumor can be removed completely, a sample of the tumor may be taken during a biopsy as a separate procedure before surgery.

See Testing Biopsy and Cytology Specimens for Cancer\(^11\) to learn more about different types of biopsies, how the biopsy samples are tested in the lab, and what the results might tell you.
Hyperlinks

2. www.cancer.org/treatment/understanding-your-diagnosis/tests/ultrasound-for-cancer.html
4. www.cancer.org/treatment/understanding-your-diagnosis/tests/mri-for-cancer.html
5. www.cancer.org/treatment/understanding-your-diagnosis/tests/imaging-radiology-tests-for-cancer.html
6. www.cancer.org/treatment/understanding-your-diagnosis/tests/understanding-your-lab-test-results.html

References


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Wilms Tumor Stages

The stage of a cancer describes how far it has spread. Your child’s treatment and prognosis (outlook) depend, to a large extent, on the cancer’s stage. Staging is based on the results of the physical exam and imaging tests (ultrasound, CT scans, etc.), which are described in Tests for Wilms Tumors, as well as on the results of surgery to remove the tumor, if it has been done.

Children’s Oncology Group (COG) staging system

A staging system is a standard way for the cancer care team to sum up the extent of the tumor. In the United States, the Children’s Oncology Group staging system is used most often to describe the extent of spread of Wilms tumors. This system divides Wilms tumors into 5 stages using Roman numerals I through V.

Stage I

The tumor is contained within one kidney and was removed completely by surgery. The tissue layer surrounding the kidney (the renal capsule) was not broken during surgery. The cancer had not grown into blood vessels in or next to the kidney. The tumor was not biopsied before surgery to remove it.

Stage II

The tumor has grown beyond the kidney, either into nearby fatty tissue or into blood vessels in or near the kidney, but it was removed completely by surgery without any apparent cancer left behind. Nearby lymph nodes (bean-sized collections of immune cells) do not contain cancer. The tumor was not biopsied before surgery.

Stage III

This stage refers to Wilms tumors that most likely have not have been removed completely. The cancer remaining after surgery is limited to the abdomen (belly). One or more of the following features may be present:

- The cancer has spread to lymph nodes in the abdomen or pelvis but not to more distant lymph nodes, such as those inside the chest.
- The cancer has grown into nearby vital structures so the surgeon could not remove it completely.
Deposits of tumor (tumor implants) are found along the inner lining of the abdominal space.
- Cancer cells are found at the edge of the sample removed by surgery, a sign that some of the cancer still remains after surgery.
- Cancer cells “spilled” into the abdominal space before or during surgery.
- The tumor was removed in more than one piece – for example, the tumor was in the kidney and in the nearby adrenal gland, which was removed separately.
- A biopsy of the tumor was done before it was removed with surgery.

**Stage IV**

The cancer has spread through the blood to organs away from the kidneys such as the lungs, liver, brain, or bones, or to lymph nodes far away from the kidneys.

**Stage V**

Tumors are found in both kidneys at the time of diagnosis.

**Tumor histology**

The other main factor in determining the prognosis and treatment for a Wilms tumor is the tumor’s histology, which is based on how the tumor cells look under a microscope. The histology can be either favorable or anaplastic. These are described in more detail in [What Are Wilms Tumors?](#).

To learn more about how the stage and histology of a Wilms tumor might affect prognosis, see [Survival Rates for Wilms Tumors](#).

**Hyperlinks**


**References**


Survival Rates for Wilms Tumors

Survival rates are often used by doctors as a standard way of discussing a person’s prognosis (outlook). These numbers tell you what portion of people in a similar situation (such as with the same type and stage of cancer) are still alive a certain amount of time after they were diagnosed. They can’t tell you exactly what will happen with any person, but they may help give you a better understanding about how likely it is that treatment will be successful. Some people find survival rates helpful, but some people might not.

For Wilms tumors, survival is often measured using a 4-year survival rate. This refers to the percentage of children who live at least 4 years after their cancer is diagnosed. For example, a 4-year survival rate of 80% means that an estimated 80 out of 100 children who have that cancer are still alive 4 years after being diagnosed. Of course, many children live much longer than 4 years (and many are cured).

To get 4-year survival rates, doctors have to look at children who were treated at least 4 years ago. Improvements in treatment since then may result in a better outlook for children now being diagnosed with Wilms tumors.

But remember, survival rates are estimates, and they can’t predict what will happen in a particular child’s case. Each child’s outlook can vary based on a number of factors specific to them. The most important factors in determining a child’s outlook are the stage and histology of the tumor. (Histology refers to how the cancer cells look under
the microscope – see [What Are Wilms Tumors?](#) But other factors can also affect a child’s outlook, such as the child’s age and how well the tumor responds to treatment.

Even when taking other factors into account, survival rates are only rough estimates. Your child’s cancer care team can tell you how the numbers below might apply, as they know your child’s situation best.

**Survival rates for Wilms tumors**

These survival rates are based on the results of the National Wilms Tumor Studies, which included most of the children treated in the United States in the last few decades. Some of these rates are based on only small numbers of children, so it’s hard to know how accurate they are.

<table>
<thead>
<tr>
<th>Tumor Stage</th>
<th>Favorable Histology</th>
<th>Focal Anaplastic</th>
<th>Diffuse Anaplastic</th>
</tr>
</thead>
<tbody>
<tr>
<td>I</td>
<td>95% - 100%</td>
<td>85% - 90%</td>
<td>75% - 80%</td>
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<tr>
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<td>85% - 90%</td>
<td>70% - 75%</td>
<td>30% - 45%</td>
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<tr>
<td>V</td>
<td>95% - 100%</td>
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<td>65% - 70%</td>
</tr>
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**Hyperlinks**


**References**


Questions to Ask Your Child’s Doctor About Wilms Tumor

It’s important to have honest, open discussions with your child’s cancer care team. Ask any question on your mind, no matter how small it might seem. Below are some questions to consider:

If the tumor has been biopsied

• What kind of kidney cancer does my child have? Is it a Wilms tumor\(^1\)?
• Is the histology\(^2\) of the tumor favorable or anaplastic?
• What is the stage of my child’s cancer, and what does that mean?
• Do we need any other tests\(^3\) before we can decide on treatment?
• How much experience do you have treating this type of cancer?
• Will we need to see other doctors?
• Who else will be on the treatment team, and what do they do?

When deciding on a treatment plan

• What are our treatment options\(^4\)?
• Are there any clinical trials\(^5\) we might want to consider?
• What do you recommend and why?
• Should we get a second opinion\(^6\)? How do we do that? Can you recommend a doctor or cancer center?
• What are the risks and side effects of the suggested treatments?
• Which side effects start shortly after treatment and which ones might develop later on?
• Will treatment affect my child’s ability to grow and develop?
• Could treatment affect my child’s ability to have children\(^7\) later on?
• Will my child have a higher long-term risk of kidney problems or other cancers?
• How soon do we need to start treatment?
• What should we do to be ready for treatment?
• How long will treatment last? What will it be like? Where will it be done?
• How will treatment affect our daily lives (school, work, etc.)?

During and after treatment

Once treatment begins, you’ll need to know what to expect and what to look for. Not all of these questions may apply, but getting answers to the ones that do may be helpful.

• How will we know if the treatment is working?
• Is there anything we can do to help manage side effects?
• What symptoms or side effects should we tell you about right away?
• How can we reach you or someone on your team on nights, weekends, or holidays?
• Who can we talk to if we have questions about costs, insurance coverage, or social support?
• What are the chances of the cancer coming back after treatment? What might our options be if this happens?
• What type of follow-up\(^8\) will my child need after treatment?

Along with these sample questions, be sure to write down any others you might have. For instance, you might want more information about recovery times so you can plan your school or work schedules. You might also want to ask about nearby or online support groups, where you may be able to get in touch with other families who have been through this.

Also keep in mind that doctors aren’t the only ones who can provide you with information. Other health care professionals\(^9\), such as nurses and social workers, may have the answers to some of your questions. You can find out more about speaking with your health care team in The Doctor-Patient Relationship\(^{10}\).

Hyperlinks

3. [www.cancer.org/treatment/understanding-your-diagnosis/tests.html](http://www.cancer.org/treatment/understanding-your-diagnosis/tests.html)

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Our team is made up of doctors and oncology certified nurses with deep knowledge of cancer care as well as journalists, editors, and translators with extensive experience in medical writing.

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Treating Wilms Tumor

If your child has been diagnosed with a Wilms tumor, your child's treatment team will discuss the options with you. It’s important to weigh the benefits of each treatment option against the possible risks and side effects.

How is Wilms tumor treated?

Overall, about 9 of 10 children with Wilms tumors are cured. A great deal of progress has been made in treating this disease in recent decades. Much of this progress is the result of children with Wilms tumors taking part in clinical trials\(^\text{1}\) of new treatments.

Today, most children with this cancer are treated in a clinical trial to try to improve on what doctors believe is the best treatment. The goal of these studies is to find ways to cure as many children as possible while limiting side effects by giving as little treatment as needed.

Most children with Wilms tumors will get more than one type of treatment. The main types of treatment are:

- Surgery for Wilms Tumors
- Chemotherapy for Wilms Tumors
- Radiation Therapy for Wilms Tumors

Common treatment approaches

In the United States, surgery is the first treatment for most Wilms tumors. In Europe, doctors often prefer to give a short course of chemotherapy before the surgery. There seems to be no difference in the results from these approaches.

The first goal of treatment is to remove the primary (main) tumor, even if the cancer has
spread to other parts of the body. Sometimes the tumor might be hard to remove because it's very large, it has spread into nearby blood vessels or other vital structures, or it’s in both kidneys. For children with these tumors, doctors might use chemotherapy, radiation therapy, or a combination of these to try to shrink the tumor(s) before surgery.

If any cancer is left after surgery, radiation therapy or more surgery may be needed.

- **Treatment by Type and Stage of Wilms Tumor**

**Who treats Wilms tumors?**

Because Wilms tumors are rare, few doctors outside of those in children’s cancer centers have much experience in treating them. Children with Wilms tumors are typically treated by a team of specialists. The doctors on this team often include:

- A **pediatric surgeon** or **pediatric urologist** (a surgeon who treats urinary system problems in children [and genital problems in boys])
- A **pediatric oncologist** (a doctor who uses chemotherapy and other medicines to treat childhood cancers)
- A **pediatric radiation oncologist** (a doctor who uses radiation therapy to treat cancer in children)

Many other specialists may be involved in your child’s care as well, including other doctors, nurses, nurse practitioners (NPs), physician assistants (PAs), psychologists, social workers, rehabilitation specialists, and other health professionals.

- [How to Find the Best Cancer Treatment for Your Child](#)
- [Navigating the Health Care System When Your Child Has Cancer](#)

**Making treatment decisions**

The treatment for Wilms tumors can often be effective, but it can also cause serious side effects. It’s important to discuss all treatment options as well as their possible side effects with your child’s doctors so you can make an informed decision.

The treatment team will also help you take care of side effects and can help you work closely with nutritionists, psychologists, social workers, and other professionals to understand and deal with medical problems, stress, and other issues related to the treatment.
If time allows, getting a second opinion from another doctor experienced with your child’s type of cancer is often a good idea. This can give you more information and help you feel more confident about the treatment plan you choose. If you aren’t sure where to go for a second opinion, ask your doctor for help.

- Questions to Ask Your Child’s Doctor About Wilms Tumor
- How to Talk to Your Child’s Cancer Care Team
- Seeking a Second Opinion

Thinking about taking part in a clinical trial

Today, most children and teens with cancer are treated at specialized children’s cancer centers. These centers offer the most up-to-date-treatment by conducting clinical trials (studies of promising new therapies). Children’s cancer centers often conduct many clinical trials at any one time, and in fact most children treated at these centers take part in a clinical trial as part of their treatment.

Clinical trials are one way to get state-of-the art cancer treatment. Sometimes they may be the only way to get access to newer treatments (although there is no guarantee that newer treatments will be better). They are also the best way for doctors to learn better methods to treat these cancers. Still, they might not be right for everyone.

If you would like to learn more about clinical trials that might be right for your child, start by asking the treatment team if your clinic or hospital conducts clinical trials.

- Clinical Trials

Considering complementary and alternative methods

You may hear about alternative or complementary methods that your doctor hasn’t mentioned to treat your child's tumor or relieve symptoms. These methods can include vitamins, herbs, and special diets, or other methods such as acupuncture or massage, to name a few.

Complementary methods refer to treatments that are used along with your regular medical care. Alternative treatments are used instead of standard medical treatment. Although some of these methods might be helpful in relieving symptoms or helping people feel better, many have not been proven to work. Some might even be harmful.

Be sure to talk to your child's cancer care team about any method you are thinking about using. They can help you learn what is known (or not known) about the method,
which can help you make an informed decision.

- Complementary and Alternative Medicine\(^8\)

**Preparing for treatment**

Before treatment, the doctors and other members of the team will help you, as a parent, understand the tests that will need to be done. The team’s social worker will also counsel you about the problems you and your child might have during and after treatments such as surgery, and might be able to help you find housing and financial aid if needed.

- When Your Child Has Cancer\(^9\)

**Help getting through cancer treatment**

Your child’s cancer care team will be your first source of information and support, but there are other resources for help when you need it. Hospital- or clinic-based support services can also be an important part of your care. These might include nursing or social work services, financial aid, nutritional advice, rehab, or spiritual help. For children and teens with cancer and their families, other specialists can be an important part of care as well.

The American Cancer Society also has programs and services – including rides to treatment, lodging, and more – to help you get through treatment. Call our National Cancer Information Center at 1-800-227-2345 and speak with one of our trained specialists.

- Finding Help and Support When Your Child Has Cancer\(^10\)
  - Find Support Programs and Services in Your Area\(^11\)

*The treatment information given here is not official policy of the American Cancer Society and is not intended as medical advice to replace the expertise and judgment of your cancer care team. It is intended to help you and your family make informed decisions, together with your doctor. Your doctor may have reasons for suggesting a treatment plan different from these general treatment options. Don’t hesitate to ask him or her questions about your treatment options.*
**Surgery for Wilms Tumors**

Surgery is the main treatment for nearly all children with Wilms tumors. It's important that it is done by a surgeon who specializes in operating on children and has experience in treating these cancers.

**Removing the tumor**

The main goal of surgery is to remove the entire Wilms tumor in one piece, if possible. This is to keep the cancer cells from spreading in the abdomen (belly). Surgeons who operate on these tumors are careful to limit the chance of this type of cancer spread whenever possible. If the surgeon finds (either with imaging tests done before surgery, or when starting the operation) that the entire tumor can’t be removed safely, other treatments may be used first. If these treatments shrink the tumor enough, surgery can then be done more safely.

Depending on the situation, different operations might be used.

**Radical nephrectomy**

A radical nephrectomy removes the entire kidney and some nearby structures. This is the most common surgery for a Wilms tumor that’s only in one kidney, as it provides the best chance of making sure all of the tumor is removed.

During this operation, the surgeon makes an incision (cut), usually down the middle of the belly, and removes the cancer along with the whole kidney, the adrenal gland that sits on top of the kidney, the surrounding fatty tissue, and the ureter (tube that carries urine from the kidney to the bladder). Most children do very well with only one kidney.

**Partial nephrectomy (nephron-sparing surgery)**

A partial nephrectomy removes only part of the kidney(s). For the small number of children who have Wilms tumors in both kidneys, this surgery might be done to try to save some normal kidney tissue. The surgeon may do a radical nephrectomy to remove the kidney containing the most tumor, and then a partial nephrectomy on the other kidney, removing just the tumor and a margin of normal kidney around it. Another option might be to do partial nephrectomies on both kidneys.

Sometimes, both kidneys need to be removed completely. The child would then need dialysis several times a week. In this procedure, a machine does the job of the kidneys.
by filtering waste products out of the blood. Once the child is healthy enough, and if a donor kidney becomes available, a kidney transplant may be an option.

Assessing the extent of the disease (surgical exploration)

When either radical or partial nephrectomy is done, another main goal of surgery is to determine the extent of the cancer and whether or not it can all be removed. Lymph nodes near the kidney will be removed during surgery to look for cancer cells in them. Cancer often spreads to lymph nodes (bean-sized collections of immune cells). Lymph node removal is known as a regional lymphadenectomy.

The other kidney and nearby organs such as the liver may also be looked at closely, and any suspicious areas biopsied (samples taken to be checked for cancer under a microscope).

Knowing if a Wilms tumor has spread to the lymph nodes, the other kidney, or other nearby organs is important in determining its stage and further treatment options.

Placing a central venous catheter (port)

Often, if the child is going to get chemotherapy, a surgeon will insert a small tube (called a central venous catheter, venous access device, or port) into a large blood vessel – usually under the collar bone. This might be done during the surgery to remove the tumor, or as a separate operation (especially if chemo is going to be given before the surgery).

One end of the catheter stays outside of the body or just under the skin, and can be used to give chemo or take blood samples without the need for more needle sticks into veins. The catheter can stay in place for months.

Possible risks and side effects of surgery

Surgery to remove a Wilms tumor is a serious operation, and surgeons are very careful to try to limit any problems either during or after surgery. Complications during surgery, such as bleeding, injuries to major blood vessels or other organs, or reactions to anesthesia, are rare, but they can happen.

Almost all children will have some pain for a while after the operation, although this can usually be helped with medicines if needed. Other problems after surgery are not common but can include internal bleeding, blood clots, infections, or problems with food
moving through the intestines.

Most children do well when only one kidney is removed. But if there are tumors in both kidneys, another concern is the loss of kidney function. In these cases, doctors must balance between making sure the tumors are removed completely and removing only as much of the kidney(s) as is needed. Children who have all or parts of both kidneys removed may need dialysis, and may eventually need a kidney transplant.

**More information about Surgery**

For more general information about surgery as a treatment for cancer, see [Cancer Surgery](#).

To learn about some of the side effects listed here and how to manage them, see [Managing Cancer-related Side Effects](#).

**Hyperlinks**


**References**


Chemotherapy for Wilms Tumors

Chemotherapy (chemo) is the use of drugs to treat cancer. These drugs enter the blood and reach all areas of the body, which makes this treatment useful for cancer that has spread or might have spread beyond the kidney.

When might chemo be used?

Most children with Wilms tumors will get chemo at some point during their treatment. (Some children with very low risk tumors might not need it.)

In the United States, chemo is usually given after surgery. Sometimes it may be needed before surgery to shrink a tumor to make the operation possible. In Europe, chemo is given before surgery and continued afterward. In both cases, the type and amount of chemo depend on the stage\(^1\) and histology\(^2\) of the cancer.

Chemo drugs used to treat Wilms tumors

A combination of chemo drugs is used to treat children with Wilms tumors. The chemo drugs used most often are:

- Actinomycin D (dactinomycin)
- Vincristine

For tumors at more advanced stages, those with anaplastic histology, or tumors that recur (come back) after treatment, other drugs might also be used, such as:

- Doxorubicin (Adriamycin)
- Cyclophosphamide
Etoposide
- Irinotecan
- Carboplatin

How is chemo given?

Chemo drugs for Wilms tumors are injected into the blood, either through a vein (IV) or through a central venous catheter (a thin tube inserted into a large blood vessel during surgery).

Different drugs, doses, and lengths of treatment are used, depending on the type and stage of the Wilms tumor and the child’s age. Most often, the drugs are given once a week for at least several months.

Chemo is usually given by a nurse in the doctor’s office or in the outpatient section of the hospital. Some children with Wilms tumors might need to stay in the hospital while they are getting chemo, but usually this is not needed.

Possible side effects of chemo

Chemo drugs can affect cells other than cancer cells, which can lead to side effects.

The side effects of chemo depend on the types and doses of drugs used, and the length of treatment. Possible short-term side effects can include:

- Hair loss
- Mouth sores
- Loss of appetite
- Nausea and vomiting
- Diarrhea or constipation
- Increased chance of infections (from having too few white blood cells)
- Easy bruising or bleeding (from having too few blood platelets)
- Fatigue or extreme tiredness (from having too few red blood cells)

Your child’s doctor and treating team will watch closely for any side effects that develop. There are often ways to lessen these side effects. For example, drugs can be given to help prevent or reduce nausea and vomiting. Be sure to ask your child’s doctor or nurse about medicines to help reduce side effects, and let them know if your child has side
effects so they can be managed.

Along with the effects listed above, some drugs can have specific side effects. For example:

- **Vincristine** can damage nerves. Some patients may have tingling, numbness, weakness, or pain, particularly in the hands and feet. (This is called *peripheral neuropathy*.)
- **Doxorubicin** can damage the heart. The risk of this happening goes up as the total amount of the drug given goes up. Doctors try to limit this risk as much as possible by not giving more than the recommended doses and by checking the heart with a test called an *echocardiogram* (an ultrasound of the heart) during treatment.
- **Cyclophosphamide** can damage the bladder, which can cause blood in the urine. The risk of this can be lowered by giving the drug with plenty of fluids and with a drug called mesna, which helps protect the bladder.

**Lab tests to check for chemo side effects**

Before each chemo session, your child’s doctor will get blood tests\(^4\) to check blood cell levels and to see how well the liver and kidneys are working. If there are problems, chemo might need to be delayed or the doses reduced.

**Long-term side effects of chemo**

Possible long-term effects of treatment are one of the major challenges children might face after cancer treatment. For example:

- If your child is given doxorubicin (Adriamycin), there is a chance it could damage the heart. Your child’s doctor will carefully watch the doses used and will check your child’s heart function with imaging tests.
- Some chemo drugs can increase the risk of developing a second type of cancer\(^5\) (such as leukemia) years after the Wilms tumor is cured. But this small increase in risk has to be weighed against the importance of chemo in treating Wilms tumor.
- Some drugs might also affect **fertility**\(^6\) (the ability to have children) years later.

See [Living as a Wilms Tumor Survivor\(^7\)] for more on the possible long-term effects of treatment.
More information about chemotherapy

For more general information about how chemotherapy is used to treat cancer, see Chemotherapy. To learn about some of the side effects listed here and how to manage them, see Managing Cancer-related Side Effects.

Hyperlinks

2. www.cancer.org/cancer/wilms-tumor/about/what-is-wilms-tumor.html
4. www.cancer.org/treatment/understanding-your-diagnosis/tests/understanding-your-lab-test-results.html

References


Radiation Therapy for Wilms Tumors

Radiation therapy uses high-energy rays or particles to kill cancer cells.

When might radiation therapy be used?

Radiation is often part of treatment for more advanced Wilms tumors (stages I, II, III, IV, and V), as well as for some earlier stage tumors with anaplastic histology. It might be used:

- After surgery to try to make sure all of the cancer is gone
- Before surgery to try to shrink the tumor to make it easier to remove
- Instead of surgery if it can't be done for some reason

For more on this, see Treatment by Type and Stage of Wilms Tumor.

How radiation therapy is done

The type of radiation used for Wilms tumors is called external beam radiation therapy. Radiation from a source outside the body is focused onto the cancer.

Before treatments start, the radiation team will take careful measurements with imaging tests such as CT or MRI scans to determine the correct angles for aiming the radiation beams and the proper dose of radiation. This planning session is called simulation. Your child may be fitted with a plastic mold that looks like a body cast to keep him or her in the same position during each treatment so that the radiation can be aimed more accurately.

Radiation is usually given 5 days a week for several weeks. Each treatment is much like getting an x-ray, although the dose of radiation is much stronger. For each session, your child lies on a special table while a machine delivers the radiation from precise angles. Each session lasts about 15 to 30 minutes, with most of the time being spent making sure the radiation is aimed correctly. The actual treatment time is much shorter. The
treatment is not painful, but some younger children may be given medicine to make them drowsy or asleep before each treatment to help make sure they stay still.

**Types of radiation therapy**

Modern radiation therapy techniques help doctors aim the treatment at the tumor more accurately than in the past. These techniques may help increase success rates and reduce side effects.

**Three-dimensional conformal radiation therapy (3D-CRT):** 3D-CRT uses the results of imaging tests such as MRI and special computers to precisely map the location of the tumor. Radiation beams are then shaped and aimed at the tumor from different directions. Each beam alone is fairly weak, which makes it less likely to damage normal body tissues, but the beams converge at the tumor to give a higher dose of radiation there.

**Intensity modulated radiation therapy (IMRT):** IMRT is an advanced form of 3D therapy. Along with shaping the beams and aiming them at the tumor from several angles, the intensity (strength) of the beams can be adjusted to limit the dose reaching the nearby normal tissues. This lets doctors deliver a higher dose to the tumor. Many major hospitals and cancer centers now use IMRT.

**Possible side effects of radiation therapy**

Radiation is often an important part of treatment, but young children’s bodies are very sensitive to it, so doctors try to use as little as possible to help avoid or limit any problems. Radiation therapy can cause both short-term and long-term side effects, which depend on the dose of radiation and where it’s aimed.

Possible **short-term** effects include:

- Effects on areas of skin that get radiation can range from mild sunburn-like changes and hair loss to more severe skin reactions.
- Radiation to the abdomen (belly) can cause nausea or diarrhea.
- Radiation therapy can make a child tired, especially after several days or weeks of treatment.

Possible **long-term** effects include:

- Radiation to the kidney area can damage the kidneys. This is more likely to be a
concern in children who need treatment in both kidneys.

- Radiation can slow the growth of normal body tissues (such as bones) that get radiation, especially in younger children. In the past this led to problems such as short bones or a curving of the spine, but this is less likely with the lower doses of radiation used today.
- Radiation to the chest area can affect the heart and lungs. This doesn’t usually cause problems right away, but in some children it might lead to heart or lung problems as they get older.
- In girls, radiation to the abdomen (belly) may damage the ovaries. This might lead to abnormal menstrual cycles or problems getting pregnant or having children later on.
- Radiation slightly increases the risk of developing a second cancer in the area, usually many years after it is given. This doesn’t happen often with Wilms tumors because the amount of radiation used is low.


**More information about radiation therapy**

To learn more about how radiation is used to treat cancer, see Radiation Therapy[^6].

To learn about some of the side effects listed here and how to manage them, see Managing Cancer-related Side Effects[^7].

**Hyperlinks**

4. [www.cancer.org/treatment/understanding-your-diagnosis/tests/mri-for-cancer.html](http://www.cancer.org/treatment/understanding-your-diagnosis/tests/mri-for-cancer.html)
References


Last Medical Review: October 17, 2018 Last Revised: October 17, 2018

Treatment by Type and Stage of Wilms Tumor

In the United States, most children with Wilms tumors are treated in clinical trials developed by the Children’s Oncology Group. The goal of these studies is to cure as many children as possible while limiting side effects by giving as little treatment as is necessary. This is done by comparing the current best treatment with one the doctors think might be better. Because of this, treatment may differ slightly from what is described here.

Treatment for Wilms tumor is based mainly on the stage of the cancer and whether its histology (how it looks under the microscope) is favorable or anaplastic. Other factors can influence treatment as well, including:

- The child’s age
- If the tumor cells have certain chromosome changes
- The size of the main tumor

In the United States, doctors prefer to use surgery as the first treatment in most cases,
and then give chemotherapy (and possibly radiation therapy) afterward. In Europe, doctors prefer to start the chemotherapy before surgery. The results from these approaches seem to be about the same.

Most often, the stage and histology of the cancer are actually determined when surgery is done to remove the cancer, because the true extent of the tumor often can’t be determined by imaging tests\(^5\) alone. The doctors use what they learn during surgery to guide further treatment. But sometimes it’s clear that the cancer has already spread beyond the kidney even before surgery is done, based on imaging tests. This can affect the order in which treatments are given, as well as the extent of surgery.

### Stage I

These tumors are only in the kidney, and surgery has completely removed the tumor along with the entire kidney, nearby structures, and some nearby lymph nodes\(^6\).

**Favorable histology:** Children younger than 2 years with small tumors (weighing less than 550 grams) may not need further treatment after surgery. But they need to be watched closely because the chance the cancer will come back is slightly higher than if they also got chemo. If the cancer does come back, the chemo drugs actinomycin D (dactinomycin) and vincristine (and possibly more surgery) are very likely to be effective at this point.

For children older than 2 and for those of any age who have larger tumors, surgery is usually followed by chemo for several months, with the drugs actinomycin D and vincristine. If the tumor cells have certain chromosome changes, the drug doxorubicin (Adriamycin) may be given as well.

**Anaplastic histology:** For children of any age who have tumors with anaplastic histology, surgery is usually followed by radiation therapy to the area of the tumor, along with chemo with actinomycin D, vincristine, and possibly doxorubicin (Adriamycin) for several months.

### Stage II

These tumors have grown outside the kidney into nearby tissues, but surgery has removed all visible signs of cancer.

**Favorable histology:** After surgery, standard treatment is chemo with actinomycin D and vincristine. If the tumor cells have certain chromosome changes, the drug doxorubicin (Adriamycin) may be given as well. The chemo is given for several months.
Anaplastic histology, with focal (only a little) anaplasia: When the child recovers from surgery, radiation therapy is given over several weeks. When this is finished, chemo (doxorubicin, actinomycin D, and vincristine) is given for about 6 months.

Anaplastic histology, with diffuse (widespread) anaplasia: After surgery, these children get radiation over several weeks. This is followed by a more intense type of chemo using the drugs vincristine, doxorubicin, etoposide, cyclophosphamide, and carboplatin, along with mesna (a drug that helps protect the bladder from the effects of cyclophosphamide), which is given for about 6 months.

Stage III

Surgery cannot remove these tumors completely because of their size or location or for other reasons. In some cases, surgery may be postponed until other treatments are able to shrink the tumor first (see below).

Favorable histology: Treatment is usually surgery if it can be done, followed by radiation therapy over several days. This is followed by chemo with 3 drugs (actinomycin D, vincristine, and doxorubicin). If the tumor cells have certain chromosome changes, the drugs cyclophosphamide and etoposide may be given as well. Chemo is given for about 6 months.

Anaplastic histology, with focal (only a little) anaplasia: Treatment starts with surgery if it can be done, followed by radiation therapy over several weeks. This is followed by chemo, usually with 3 drugs (actinomycin D, vincristine, and doxorubicin) for about 6 months.

Anaplastic histology, with diffuse (widespread) anaplasia: Treatment starts with surgery if it can be done, followed by radiation therapy over several weeks. This is followed by chemo, usually with the drugs vincristine, doxorubicin, etoposide, cyclophosphamide, and carboplatin, along with mesna (a drug that helps protect the bladder from the effects of cyclophosphamide). Chemo lasts about 6 months.

In some instances the tumor may be very large or may have grown into nearby blood vessels or other structures so that it can’t be removed safely. In these children, a small biopsy sample is taken from the tumor to be sure that it’s a Wilms tumor and to determine its histology. Then chemo is started. Usually the tumor will shrink enough within several weeks so that surgery can be done. If not, then radiation therapy might be given as well. Chemo will be started again after surgery. If radiation was not given before surgery, it’s given after surgery.
Stage IV

These tumors have already spread to distant parts of the body at the time of diagnosis. As with stage III tumors, surgery to remove the tumor might be the first treatment, but it might need to be delayed until other treatments can shrink the tumor (see below).

**Favorable histology:** Surgery to remove the tumor is the first treatment if it can be done, followed by radiation therapy. The entire abdomen will be treated if there is still some cancer left after surgery. If the cancer has spread to the lungs, low doses of radiation might also be given to that area. This is followed by chemo, usually with 3 drugs (actinomycin D, vincristine, and doxorubicin) for about 6 months. If the tumor cells have certain chromosome changes, the drugs cyclophosphamide and etoposide may be given as well.

**Anaplastic histology:** Treatment might start with surgery if it can be done, followed by radiation therapy. The entire abdomen will be treated if there is still some cancer left after surgery. Low doses of radiation will also be given to the lungs if the cancer has spread there. This is followed by chemo with the drugs vincristine, doxorubicin, etoposide, cyclophosphamide, and carboplatin, along with mesna given for about 6 months. If the tumor cells have diffuse (widespread) anaplasia, some doctors might try the chemo drugs irinotecan and vincristine first instead (although this is not yet a commonly used treatment). The treatment would then be adjusted if the tumor shrinks in response to these drugs.

If the tumor is too large or has grown too much to be removed safely with surgery first, a small biopsy sample may be taken from the tumor to be sure that it’s a Wilms tumor and to determine its histology. Chemo and/or radiation therapy may then be used to shrink the tumor. Surgery might be an option at this point. This would be followed by more chemo and radiation therapy if it wasn’t given already.

For stage IV cancers that have spread to the liver, surgery may be an option to remove any liver tumors that still remain after chemo and radiation therapy.

Stage V

Treatment for children with tumors in both kidneys is unique for each child, although it typically includes surgery, chemo, and radiation therapy at some point.

**Biopsies** (tissue samples) of tumors in both kidneys and of nearby lymph nodes may be taken first, although not all doctors feel this is needed because when both kidneys have tumors, the chance that they are Wilms tumors is very high.
Chemo is typically given first to try to shrink the tumors. The drugs used will depend on the extent and histology (if known) of the tumors. After about 6 weeks of chemo, surgery (partial nephrectomy) may be done to remove the tumors if enough normal kidney tissue can be left behind. If the tumors haven’t shrunk enough, treatment may include more chemo or radiation therapy for about another 6 weeks. Surgery (either partial or radical nephrectomy) may then be done. This is followed by more chemo, possibly along with radiation therapy if it hasn’t been given already.

If not enough functioning kidney tissue is left after surgery, a child may need dialysis, a procedure where a special machine filters waste products out of the blood several times a week. If there is no evidence of any cancer after a year or two, a donor kidney transplant may be done.

**Recurrent Wilms tumor**

The prognosis and treatment for children with Wilms tumor that recurs (comes back after treatment) depends on their prior treatment, the cancer’s histology (favorable or anaplastic), and where it recurs. The outlook is generally better for recurrent Wilms tumors with the following features:

- Favorable histology
- Initial diagnosis of stage I or II
- Initial chemo with vincristine and actinomycin D only
- No previous radiation therapy

The usual treatment for these children is surgery to remove the recurrent cancer (if possible), radiation therapy (if not already given to the area), and chemo, often with drugs different from those used during first treatment.

Recurrent Wilms tumors that do not have the features above are much harder to treat. These children are usually treated with aggressive chemo, such as the ICE regimen (ifosfamide, carboplatin, and etoposide) or others being studied in clinical trials. Very high-dose chemo followed by a stem cell transplant (sometimes called a bone marrow transplant) might also be an option in this situation, although this is still being studied.

*The treatment information given here is not official policy of the American Cancer Society and is not intended as medical advice to replace the expertise and judgment of your cancer care team. It is intended to help you and your family make informed decisions, together with your doctor. Your doctor may have reasons for suggesting a treatment plan different from these general treatment options. Don’t hesitate to ask him or her questions about your treatment options.*
Hyperlinks

2. https://childrensoncologygroup.org/
5. www.cancer.org/treatment/understanding-your-diagnosis/tests.html

References


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The American Cancer Society medical and editorial content team (www.cancer.org/cancer/acscase-medical-content-and-news-staff.html)
Our team is made up of doctors and oncology certified nurses with deep knowledge of cancer care as well as journalists, editors, and translators with extensive experience in medical writing.

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After Wilms Tumor Treatment

Living as a Wilms Tumor Survivor

For many people, cancer treatment often raises questions about next steps as a survivor.

- What Happens After Treatment for Wilms Tumor?

What Happens After Treatment for Wilms Tumor?

During treatment for Wilms tumors, the main concerns for most families are the daily aspects of getting through treatment and beating the cancer. After treatment, the concerns tend to shift toward the short- and long-term effects of the cancer and its treatment, and concerns about the cancer coming back.

It’s certainly normal to want to put the tumor and its treatment behind you, and to get back to a life that doesn’t revolve around cancer. But it’s important to realize that follow-up care is a central part of treatment that offers your child the best chance for long-term recovery.

Follow-up exams and tests

Your child’s health care team will discuss a follow-up schedule with you, which will
include physical exams and imaging tests\(^2\) (such as chest x-rays, ultrasounds, and CT scans) to look for the growth or return of the tumor, or any problems related to treatment.

If your child had parts or all of a kidney removed, blood and urine tests will be done to check how well the remaining kidney tissue is working. If your child received the drug doxorubicin (Adriamycin) during chemotherapy\(^3\), the doctor may also order tests to check the function of your child’s heart.

The recommended schedule for follow-up exams and tests depends on several factors, including:

- The initial stage\(^4\) and histology\(^5\) (favorable or anaplastic) of the tumor
- If the child has a genetic syndrome\(^6\) related to the tumor
- The type of treatment the child received
- Any problems that the child may have had during treatment

Doctor visits and tests will be more frequent at first (about every 6 to 12 weeks for the first couple of years), but the time between visits may be extended as time goes on.

During this time, it’s important to report any new symptoms to your child’s doctor right away, so that the cause can be found and treated, if needed. Your child’s doctor can give you an idea of what to watch for.

If the tumor does come back\(^7\), or if it doesn’t respond to treatment, your child’s doctors will discuss the treatment options with you.

Children with bilateral Wilms tumors (tumors in both kidneys) or Denys-Drash syndrome\(^8\) will also need regular tests to look for possible early signs of kidney failure (including urine tests, blood pressure checks, and blood tests of kidney function).

**Ask the cancer care team for a survivorship care plan**

Talk with the treatment team about developing a survivorship care plan\(^9\). This plan might include:

- A summary of the diagnosis, tests done, and treatment given
- A suggested schedule for follow-up exams and tests
- A schedule for other tests that might be needed in the future, such as early detection (screening) tests for other types of cancer, or tests to look for long-term
health effects from the cancer or its treatment
• A list of possible late- or long-term side effects from treatment, including what to watch for and when to contact the doctor

Keeping health insurance and copies of medical records

As much as you might want to put the experience behind you once treatment is completed, it’s also very important to keep good records of your child’s medical care during this time. Gathering these details soon after treatment may be easier than trying to get them at some point in the future. This can be very helpful later on if your child changes doctors. Learn more about this in Keeping Copies of Important Medical Records. It’s also very important to keep health insurance coverage. Tests and doctor visits can cost a lot, and even though no one wants to think of the tumor coming back, this could happen.

Possible late and long-term effects of treatment

Because of major advances in treatment, most children treated for Wilms tumor are now surviving into adulthood. Doctors have learned that treatment can affect children’s health later in life, so watching for health effects as they get older has become more of a concern in recent years.

Just as the treatment of childhood cancer requires a very specialized approach, so does the care and follow-up after treatment. The earlier any problems can be recognized, the more likely it is they can be treated effectively. Young people treated for Wilms tumor are at risk, to some degree, for several possible late effects of their cancer treatment. It’s important to discuss what these possible effects might be with your child’s medical team so you know what to watch for and report to the doctor.

The risk of late effects depends on a number of factors, such as the specific treatments the child had, the doses of treatment, and the age of the child when being treated. These late effects may include:

• Reduced kidney function
• Heart or lung problems after getting certain chemotherapy drugs or radiation therapy to the chest
• Slowed or delayed growth and development
Changes in sexual development and ability to have children\textsuperscript{15}, especially in girls
- Increased risk of second cancers\textsuperscript{16} later in life (although these are rare)

There may be other possible complications from treatment as well. Your child’s doctors should discuss any possible problems with you.

**Long-term follow-up care**

To help increase awareness of late effects and improve follow-up care of childhood cancer survivors throughout their lives, the Children’s Oncology Group (COG)\textsuperscript{17} has developed long-term follow-up guidelines for survivors of childhood cancers. These guidelines can help you know what to watch for, what type of screening tests should be done to look for problems, and how late effects can be treated.

It’s very important to discuss possible long-term complications with your child’s health care team, and to make sure there’s a plan in place to watch for these problems and treat them, if needed. To learn more, ask your child’s doctors about the COG survivor guidelines. You can also read them online at www.survivorshipguidelines.org\textsuperscript{18}. The guidelines themselves are written for health care professionals. Patient versions of some of the guidelines are available (as “Health Links”) on the site as well, but we urge you to discuss them with your doctor.

For more about some of the possible long-term effects of treatment, see Late Effects of Childhood Cancer Treatment\textsuperscript{19}.

**Emotional and social issues**

Most children with Wilms tumors are very young when they are diagnosed. Still, some children may have emotional or psychological issues\textsuperscript{20} that need to be addressed during and after treatment. Depending on their age, they may also have some problems with normal functioning and school work.

These types of issues can often be helped with support and encouragement. Doctors and other members of the health care team can also often recommend special support programs and services to help children after treatment. For more information, see When Your Child’s Treatment Ends\textsuperscript{21}.

Parents and other family members can also be affected, both emotionally and in other ways\textsuperscript{22}. Some common family concerns during treatment include financial stresses, traveling to and staying near the cancer center/hospital, the possible loss of a job, and
the need for home schooling. Social workers and other professionals at treatment centers can help families sort through these issues.

Centers that treat many patients with Wilms tumors may have programs to introduce new patients and their families to others who have finished their treatment. This can give parents an idea of what to expect during and after treatment, which is very important.

Support groups for families of children with cancer can also be helpful. If you need help finding such a group, call your American Cancer Society at 1-800-227-2345 and we can put you in touch with a group or resource that may work for you.

Hyperlinks

2. [www.cancer.org/treatment/understanding-your-diagnosis/tests.html](http://www.cancer.org/treatment/understanding-your-diagnosis/tests.html)
17. [https://childrensoncologygroup.org/](https://childrensoncologygroup.org/)
18. [http://www.survivorshipguidelines.org](http://www.survivorshipguidelines.org)
23. www.cancer.org/treatment/support-programs-and-services.html

References


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