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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?](#)
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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?](#)¹ For information

about the differences between childhood cancers and adult cancers, see [Cancer in Children²](#).

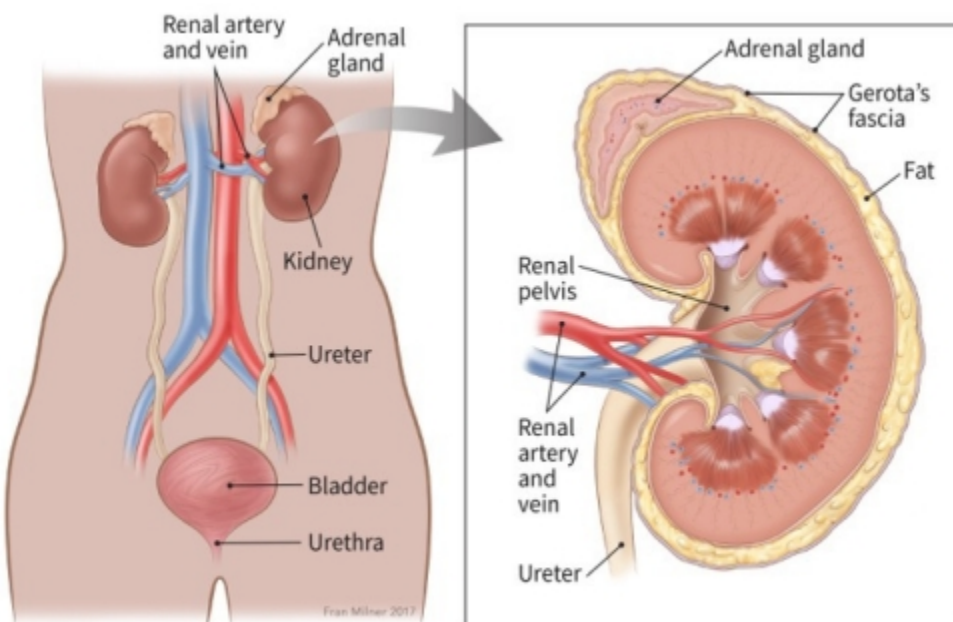
About the kidneys

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](#)³, but sometimes [chemotherapy](#)⁴ 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](#)⁵. It's usually much like the intensive treatment used for Wilms tumors with anaplastic histology (see [Treatment of Wilms Tumors by Type and Stage](#)⁶).

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](#)⁷, and usually includes [chemotherapy](#)⁸ with several different drugs.

Renal cell carcinoma

This is the most common type of [kidney cancer in adults](#)⁹, 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](#)¹⁰ 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

1. www.cancer.org/cancer/cancer-basics/what-is-cancer.html
2. www.cancer.org/cancer/cancer-in-children.html
3. www.cancer.org/treatment/treatments-and-side-effects/treatment-types/surgery.html
4. www.cancer.org/treatment/treatments-and-side-effects/treatment-types/chemotherapy.html
5. www.cancer.org/treatment/treatments-and-side-effects/clinical-trials.html
6. www.cancer.org/cancer/wilms-tumor/treating/by-stage.html
7. www.cancer.org/treatment/treatments-and-side-effects/clinical-trials.html
8. www.cancer.org/treatment/treatments-and-side-effects/treatment-types/chemotherapy.html
9. www.cancer.org/cancer/kidney-cancer.html

10. www.cancer.org/treatment/treatments-and-side-effects/treatment-types/surgery.html

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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

1. www.cancer.org/cancer/wilms-tumor/detection-diagnosis-staging/survival-rates.html
2. <https://cancerstatisticscenter.cancer.org>

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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](#)² (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](#)³ cause Wilms tumors and affect how aggressive these tumors are likely to be.

As doctors have learned how to [treat Wilms tumors](#)⁴ 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](#)⁵ 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](#)⁶.
- 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](#)⁷ (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](#)⁸ 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

1. www.cancer.org/treatment/children-and-cancer/when-your-child-has-cancer/late-effects-of-cancer-treatment.html
2. <https://childrensoncologygroup.org/>
3. www.cancer.org/cancer/wilms-tumor/causes-risks-prevention/what-causes.html
4. www.cancer.org/cancer/wilms-tumor/treating.html
5. www.cancer.org/treatment/treatments-and-side-effects/clinical-trials.html
6. www.cancer.org/cancer/wilms-tumor/treating/surgery.html
7. www.cancer.org/treatment/treatments-and-side-effects/treatment-types/stem-cell-transplant.html

8. www.cancer.org/treatment/treatments-and-side-effects/treatment-types/targeted-therapy.html

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