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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?](#)

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

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/treatment/understanding-your-diagnosis/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

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

References

American Cancer Society. *Cancer Facts & Figures 2020*. Atlanta, Ga: American Cancer Society; 2020.

Chintagumpala M, Muscal JA. Presentation, diagnosis, and staging of Wilms tumor. UpToDate. Accessed at www.uptodate.com/contents/presentation-diagnosis-and-staging-of-wilms-tumor on August 15, 2018.

Fernandez CV, Geller JI, Ehrlich PF, et al. Chapter 29: Renal Tumors. In: Pizzo PA, Poplack DG, eds. *Principles and Practice of Pediatric Oncology*. 7th ed. Philadelphia, Pa: Lippincott Williams & Wilkins; 2016.

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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.

- [effects-of-cancer-treatment.html](#)
2. [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](#)
 9. [www.cancer.org/research/acs-research-highlights/childhood-cancer-research-highlights.html](#)

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