

About Wilms Tumor

Get an overview of Wilms tumors and the latest key statistics in the US.

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 **nephroblastoma**) is the most common type of kidney cancer in children. About 9 of 10 kidney cancers in children are Wilms tumors. These cancers are different from <u>adult kidney cancers</u>¹.

Wilms tumors and the kidneys

- How Wilms tumors form
- How Wilms tumors are grouped
- Other types of kidney cancers in children

What Is Cancer?²

Cancer starts when cells in the body begin to grow out of control. Cells in nearly any part of the body can become cancer cells. Learn more here.

Anatomy Gallery ³

Explore our 3D interactive tours of the human body.

Wilms tumors and the kidneys

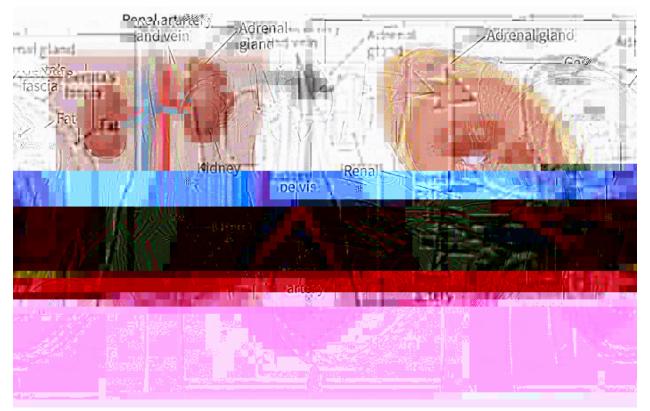
To understand Wilms tumors, it helps to know about the kidneys and how they work.

The kidneys are 2 bean-shaped organs attached to the back wall of the abdomen (belly), just to the left and right of the backbone. The job of the kidneys is to:

- Filter the blood to remove excess water, salt, and waste products, which leave the body as urine
- Help control blood pressure
- 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.

How Wilms tumors form

The kidneys develop very early as a fetus grows in the womb. Certain cells are programmed to turn into mature kidney cells. But sometimes, some of these early kidney cells don't mature like they are supposed to. Instead, they stay in the body in the form of early (not mature) kidney cells. They might stay in the body 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 could result in a Wilms tumor.

How Wilms tumors are grouped

Wilms tumors can be grouped in different ways:

given as well. These tumors sometimes come back soon after treatment, so children who've had them 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. They are also harder to cure. Because these tumors are rare, treatment is often given as part of a <u>clinical trial</u>⁶. It's usually a lot like the intensive treatment used for Wilms tumors with anaplastic histology (see <u>Treatment of Wilms Tumors by Type and Stage</u>⁷).

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. 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 <u>clinical trial</u>⁸. It usually includes <u>chemotherapy</u>⁹ with several different drugs.

Renal cell carcinoma

This is the most common type of <u>kidney cancer in adults</u>¹⁰, but it also accounts for a small number of kidney cancers in children. It's rare in young children. But in older teens, it's actually more common than Wilms tumor.

<u>Surgery</u>¹¹ to remove the kidney (or just the tumor) is the main treatment for these cancers, if it can be done. The outlook 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 I /GS 0.2 Tr4eh

- 5. <u>www.cancer.org/cancer/managing-cancer/treatment-types/chemotherapy.html</u>
- 6. <u>www.cancer.org/cancer/managing-cancer/making-treatment-decisions/clinical-</u> <u>trials.html</u>
- 7. www.cancer.org/cancer/types/wilms-tumor/treating/by-stage.html
- 8. <u>www.cancer.org/cancer/managing-cancer/making-treatment-decisions/clinical-</u> <u>trials.html</u>
- 9. <u>www.cancer.org/cancer/managing-cancer/treatment-types/chemotherapy.html</u>
- 10. www.cancer.org/cancer/types/kidney-cancer.html
- 11. www.cancer.org/cancer/managing-cancer/treatment-types/surgery.html

References

Fernandez CV, Geller JI, Ehrlich PF, et al. Chapter 24: Renal Tumors. In: Blaney SM,

Key Statistics for Wilms Tumors

(PDQ®)–Health Professional Version. 2024. Accessed at https://www.cancer.gov/types/kidney/hp/wilms-treatment-pdq on November 21, 2024.

Smith V, Chintagumpala M. Clinical presentation, diagnosis, and staging of Wilms tumor. UpToDate. 2024. Accessed at https://www.uptodate.com/contents/clinical-presentation-diagnosis-and-staging-of-wilms-tumor on November 21, 2024.

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What's New in Wilms Tumor Research?

Over the past few decades, research into Wilms tumors 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 <u>long-term side effects</u>¹ from treatment, so more research is needed.

In the United States, much of the research on Wilms tumors is coordinated by the <u>Children's Oncology Group</u>² (COG), whose main goal is to improve the treatment and quality of life for children with Wilms tumors 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.

The information below is an overview of the latest research into Wilms tumors, including genetics and treatments.

- Research into the genetics of Wilms tumors
- Research into the treatment of Wilms tumors

Research into the genetics of Wilms tumors

Researchers continue to learn how <u>changes in certain genes</u>³ inside kidney cells cause Wilms tumors and affect how aggressive these tumors are likely to be.

As doctors have learned how better to <u>treat Wilms tumors</u>⁴, they've looked for ways to determine which children might need more intense treatment to be cured, and which children might be spared from intense treatment (and its possible side effects).

standard treatments or that would otherwise have a poor outlook.

As researchers have learned more about the gene changes in cancer cells, they have developed newer drugs that target these changes. <u>Targeted drugs</u>⁸ work differently from standard chemo drugs. They sometimes work when chemo drugs don't, and they often have different (and less severe) side effects. Targeted therapies have 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.

Childhood Cancer Research Highlights 9

The American Cancer Society is committed to finding new answers to help every child and family affected by cancer--see some of our latest research.

Hyperlinks

- 1. <u>www.cancer.org/cancer/survivorship/children-with-cancer/late-effects-of-cancer-</u> <u>treatment.html</u>
- 2. <u>childrensoncologygroup.org/</u>
- 3. <u>www.cancer.org/cancer/types/wilms-tumor/causes-risks-prevention/what-causes.html</u>
- 4. <u>www.cancer.org/cancer/types/wilms-tumor/treating.html</u>
- 5. <u>www.cancer.org/cancer/managing-cancer/making-treatment-decisions/clinical-</u> <u>trials.html</u>
- 6. <u>www.cancer.org/cancer/types/wilms-tumor/treating/surgery.html</u>
- 7. <u>www.cancer.org/cancer/managing-cancer/treatment-types/stem-cell-</u> <u>transplant.html</u>
- 8. <u>www.cancer.org/cancer/managing-cancer/treatment-types/targeted-therapy.html</u>
- 9. <u>www.cancer.org/research/acs-research-highlights/childhood-cancer-research-highlights.html</u>

References

Dix DB, Seibel NL, Chi YY, et al. Treatment of stage IV favorable histology Wilms tumor with lung metastases: A report from the Children's Oncology Group AREN0533 study. *J*

Clin Oncol. 2018;36(16):1564-1570.