Wilms Tumor Causes, Risk Factors, and Prevention

Learn about the risk factors for Wilms tumors.

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 like cancer. Different cancers have different risk factors.

Lifestyle-related risk factors like body weight, physical activity, diet, and tobacco and alcohol use play a major role in many adult cancers. But these factors usually take many years to influence cancer risk. 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 risk 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
- Race/ethnicity
- Sex
- Family history of Wilms tumor
- Certain genetic syndromes/birth defects

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 Black children than in White children, and it's lower among Asian American children. The reason for this is not known.

Sex

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. These children most likely inherit chromosomes with an abnormal or missing gene[1] from a n

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 a Wilms tumor also has one or more 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 syndrome is linked with certain physical and mental issues (although not all children have all of them):

- **W**ilms tumor
- **A**niridia (complete or partial lack of the iris [colored area] of the eyes)
- **G**enitourinary tract abnormalities (defects of the kidneys, urinary tract, penis, scrotum, clitoris, testicles, or ovaries)
- **R**ange of developmental delays (including intellectual disabilities)

Children with this syndrome have about a 50% chance of developing a Wilms tumor. When they do develop Wilms tumors, it tends to happen at an earlier age, and often in both kidneys.

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?).

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 this risk is not as high as with Denys-Drash syndrome. 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 Wilms tumors (or, less often, other cancers that develop during childhood).

This syndrome is caused by a defect in chromosome 11 that affects several genes.

Other syndromes

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

- Perlman syndrome
- Fanconi anemia
- 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

1. www.cancer.org/cancer/understanding-cancer/genes-and-cancer/gene-changes.html

References

Fernandez CV, Geller JI, Ehrlich PF, et al. Chapter 24: Renal Tumors. In: Blaney SM, Adamson PC, Helman LJ, eds. *Pizzo and Poplack's Principles and Practice of Pediatric Oncology*. 8th ed. Philadelphia, Pa: Lippincott Williams & Wilkins; 2021.

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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 Causes Wilms Tumors?

It's not always clear exactly what causes Wilms tumors.

There is a clear link between Wilms tumors and certain birth defect syndromes and genetic changes[1]. But most children with this type of cancer do not have any known birth defects or inherited gene changes.

Researchers have made great progress in understanding how normal kidneys develop, as well as how this process can go wrong, leading to a Wilms tumor. Here is what we

know so far about the causes of Wilms tumors.

How Wilms tumors form

A Wilms tumor can form when early kidney cells don't mature and instead grow out of control.

The kidneys develop very early as a fetus grows in the womb. Sometimes, certain cells that are programmed to turn into kidney cells don't mature like they're 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 a child is 3 to 4 years old. But if this doesn't happen, the cells might somehow begin to grow out of control. This can result in a Wilms tumor.

How gene changes can affect kidney development

Cells are the building blocks for all life, including the human body. Our **genes** control how our cells work.

Different genes have different functions in our bodies. If they are working properly, certain genes help control when our cells grow, divide to make new cells, or repair mistakes in our DNA (the chemical in each of our cells that makes up our genes).

Genes also cause cells to die when they're supposed to. If these genes aren't working properly, it can lead to cells growing out of control.

For example:

- Changes in genes that normally help cells grow, divide, or stay alive can lead to these genes being more active than they should be, causing them to become **oncogenes**. These genes can result in cells growing out of control.
- Genes that normally help keep cell division under control or cause cells to die at the right time are known as **tumor suppressor genes**. Changes that turn off these genes can result in cells growing out of control.

- Some genes normally help repair mistakes in a cell's DNA. Changes that turn off these **DNA repair genes** can result in the buildup of DNA changes within a cell, which might lead to them growing out of control.

Any of these types of DNA changes might lead to cells growing out of control and forming a tumor. To learn more, see Oncogenes, Tumor Suppressor Genes, and DNA Repair Genes[2].

Gene changes in Wilms tumors

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 (inherited). But most Wilms tumors don't seem to be caused by inherited gene mutations. Instead, they seem to be caused by gene changes that happen early in a child's life, perhaps even before birth.

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 lower the risk of these cancers.

Hyperlinks

1. www.cancer.org/cancer/understanding-cancer/genes-and-cancer/gene-changes.html.
2. www.cancer.orgcancer/understanding-cancer/genes-and-cancer/oncogenes-tumor-suppressor-genes.html

References

Fernandez CV, Geller JI, Ehrlich PF, et al. Chapter 24: Renal Tumors. In: Blaney SM, Adamson PC, Helman LJ, eds. *Pizzo and Poplack's Principles and Practice of Pediatric Oncology*. 8th ed. Philadelphia, Pa: Lippincott Williams & Wilkins; 2021.

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Can Wilms Tumors Be Prevented?

Certain lifestyle changes (such as staying at a healthy weight or quitting smoking), can reduce the risk of many adult cancers. But at this time, there are no known ways to prevent most cancers in children. This includes Wilms tumors.