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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):

- **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)
- **M**ental **R**etardation

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

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

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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.

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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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