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 tumor.

- What Are the Risk Factors for Wilms Tumor?
- Do We Know What Causes Wilms Tumor?

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.

What Are the Risk Factors for Wilms Tumor?

A risk factor is anything that affects the 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 has not 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

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

Girls have a slightly higher risk of Wilms tumor than 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 from a parent that increases their risk of developing Wilms tumor. Surprisingly, the relative with Wilms tumor is not usually a parent.

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):

- Wilms tumor
- Aniridia (complete or partial lack of the iris [colored area] of the eyes)
- Genitourinary tract abnormalities (defects of the kidneys, urinary tract, penis, scrotum, clitoris, testicles, or ovaries)
- Mental Retardation

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 Do we know 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)

References

See all references for Wilms Tumor

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Do We Know What Causes Wilms Tumor?

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. Changes (mutations) in certain genes in early kidney cells can lead to problems as the kidneys develop. Some of the cells that are supposed to develop into mature kidney cells stay as early kidney cells instead. Clusters of these early kidney cells sometimes remain after the baby is born. Usually, these cells mature by the time the child is 3 to 4 years old. If this doesn’t happen, the cells might somehow begin to grow out of control, which might result in a Wilms tumor.

Normal human cells grow and function based mainly on the information contained in each cell’s chromosomes (long strands of DNA in each cell). DNA carries the instructions for nearly everything our cells do. DNA is divided into units called genes.

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. Others 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 that turn on oncogenes or turn off tumor suppressor genes. These gene changes can be inherited from a parent (as is sometimes the case with childhood cancers), or they may happen during a person’s lifetime as cells in the body divide to make new cells.

Changes in certain genes increase the chance that some kidney cells will remain in an early form and turn into a Wilms tumor.

For example, a small number of Wilms tumors are caused by changes (mutations) 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 the section What are the risk factors for Wilms tumor? are sometimes found with Wilms tumors.

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.
In a small number of Wilms tumors there is a change in a tumor suppressor gene known as \textit{WTX}, which is found on the X chromosome. Another gene that is sometimes altered in Wilms tumor cells is known as \textit{CTNNB1}. It’s not clear exactly what causes these genes to be altered.

Because the genes described above are not altered in all Wilms tumors, there must be gene changes that have not yet been found. In many cases, more than one gene change is involved.

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. Other 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 is important to know that there is nothing these children or their parents could have done to prevent these cancers.

- References
  See all references for Wilms Tumor

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Can Wilms Tumor 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 protect against most of these cancers. Experts think these cancers come from cells that were in the fetus but failed to develop into mature kidney cells. This doesn’t seem to be caused by anything a mother could avoid during pregnancy.
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
  See all references for Wilms Tumor

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