About Wilms Tumor

Overview and Types

If your child has been diagnosed with 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 Is Wilms Tumor?
- What Are the Differences Between Cancers in Adults and Children?

Research and Statistics

See the latest estimates for new cases of Wilms tumor in the US and what research is currently being done.

- What Are the Key Statistics About Wilms Tumor?
- What’s New in Wilms Tumor Research and Treatment?

What Are the Differences Between Cancers in Adults and Children?

Cancers that develop in children are often different from those that develop in adults. Childhood cancers are often the result of DNA changes in cells very early in life, sometimes even before birth. Unlike many cancers in adults, childhood cancers are not strongly linked to lifestyle or environmental risk factors.

There are exceptions, but childhood cancers tend to respond better to treatments such as chemotherapy. Children’s bodies also tend to tolerate chemotherapy better than adults’ bodies do. But cancer treatments such as chemotherapy and radiation therapy can have some long-term side effects, so children who have had cancer need careful attention for the rest of their lives.
Since the 1960s, most children and teens with cancer have been treated at specialized centers designed for them. These centers offer the advantage of being treated by a team of specialists who know the differences between adult and childhood cancers, as well as the unique needs of children with cancer and their families. This team usually includes pediatric oncologists (childhood cancer doctors), surgeons, radiation oncologists, pathologists, pediatric oncology nurses, and nurse practitioners. These centers also have psychologists, social workers, child life specialists, nutritionists, rehabilitation and physical therapists, and educators who can support and educate the entire family.

Most children with cancer in the United States are treated at a center that is a member of the Children's Oncology Group (COG). All of these centers are associated with a university or children's hospital. As we have learned more about treating childhood cancer, it has become even more important that treatment be given by experts in this area.

Any time a child or teen is diagnosed with cancer, it affects every family member and nearly every aspect of the family's life. You can read more about coping with all these changes in Children Diagnosed With Cancer: Dealing With Diagnosis.

References
See all references for Wilms Tumor

What Is Wilms Tumor?

Cancer starts when cells in the body begin to grow out of control. Cells in nearly any part of the body can become cancer, and can spread to other areas of the body. To learn more about how cancers start and spread, see What Is Cancer?

Wilms tumor (also called Wilms' tumor or nephroblastoma) is a type of cancer that starts in the kidneys. It is the most common type of kidney cancer in children. It is named after Max Wilms, a German doctor who wrote one of the first medical articles about the disease in 1899.

About the kidneys
The kidneys are 2 bean-shaped organs that are attached to the back wall of the abdomen (see picture). One kidney is just to the left and the other just to the right of the backbone. The lower rib cage protects the kidneys.

Small glands called adrenal glands sit on top of each of the kidneys. Each kidney and adrenal gland is surrounded by fat and a thin, fibrous capsule (known as Gerota’s fascia).

The main job of the kidneys is filtering blood coming in from the renal arteries to rid the body of excess water, salt, and waste products. These substances become urine. Urine leaves the kidneys through long, slender tubes called ureters that connect to the bladder. Urine flows down the ureters into the bladder, and is stored there until the person urinates.

The kidneys also have other jobs:

- They help control blood pressure by making a hormone called renin.
- They help make sure the body has enough red blood cells by making a hormone called erythropoietin. This hormone tells the bone marrow to make more red blood
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.

**Wilms tumors**

Wilms tumors are the most common cancers in children that start in the kidneys. About 9 of 10 kidney cancers in children are Wilms tumors.

Most Wilms tumors are *unilateral*, which means they affect only one kidney. Most often there is only one tumor, but 5% to 10% of children with Wilms tumors have more than one tumor in the same kidney. About 5% 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.

Even if a doctor thinks a child might have a cancer such as Wilms tumor based on a physical exam or imaging tests, they can’t be sure until a small piece of the tumor is checked in a lab.

**Types of Wilms tumor**

Wilms tumors are grouped into 2 major types based on how they look under a microscope (their histology):

**Favorable histology:** Although the cancer cells in these tumors don’t look quite normal, there is no anaplasia (see next paragraph). More than 9 of 10 Wilms tumors have a favorable histology. The chance of curing children with these tumors is very good.

**Unfavorable histology (anaplastic Wilms tumor):** In these tumors, the look of the cancer cells varies widely, and the cells’ nuclei (the central parts that contain the DNA) tend to be very large and distorted. This is called *anaplasia*. The more anaplasia a tumor has, the harder it is to cure.

**Other types of kidney cancers in children**
Most kidney cancers in children are Wilms tumors, but in rare cases children can develop other types of kidney tumors.

**Mesoblastic nephroma**

These tumors usually appear in the first few months of life. Children are usually cured with surgery, but sometimes chemotherapy is given as well. These tumors sometimes come back soon after treatment, so children who have had these tumors 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, and they are harder to cure. Because these tumors are rare, treatment is often given as part of a clinical trial. It’s usually much like the intensive treatment used for Wilms tumors with unfavorable histology (see Treatment of Wilms tumor by type and stage).

**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, and usually includes chemotherapy with several different drugs.

**Renal cell carcinoma**

This is the most common type of kidney cancer in adults, but it also accounts for a small number of kidney tumors in children. It’s rare in young children, but it’s actually more common than Wilms tumor in older teens.

Surgery to remove the kidney 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.

- **References**

  See all references for Wilms Tumor
What Are the Key Statistics About Wilms Tumor?

Each year, about 500 new cases of Wilms tumors are diagnosed in the United States. This number has been fairly stable for many years. About 5% of all cancers in children are Wilms tumors.

Wilms tumors tend to occur in young children. The average age at diagnosis is about 3 to 4 years. It becomes less common as children grow older and is uncommon after age 6. It’s very rare in adults, although cases have been reported.

Statistics related to survival for Wilms tumors are discussed in the section “Survival rates for Wilms tumor, by stage and histology.”

Visit the American Cancer Society’s Cancer Statistics Center for more key statistics.

- References
See all references for Wilms Tumor

What’s New in Wilms Tumor Research and Treatment?

Over the past few decades, research into Wilms tumor has led to great 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 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 (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 tumor.

**Biology of Wilms tumors**

Research is continuing to unravel how changes in certain genes cause Wilms tumors and affect how aggressive tumors are likely to be.

As doctors have learned how to treat Wilms tumors more effectively, they have begun to look for ways to determine which children might be spared from more intense treatment and which children might need more aggressive treatment to be cured. For example, recent 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.

Earlier studies found treatments that were very effective in curing Wilms tumors with favorable histology. Current clinical trials 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 with very favorable outlooks need any treatment other than surgery. Recent studies from Europe have suggested that in some cases chemo may not need to be continued as long as previously thought.

The outlook for children with Wilms tumors with unfavorable 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, 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 Wilms tumor cells, they have started to develop newer drugs that specifically target these changes. Targeted drugs 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 adult cancers.

Because Wilms tumors with favorable histology are usually cured with surgery and chemotherapy, and because Wilms tumors with unfavorable histology are uncommon, most research on targeted drugs so far has been done on cells growing in lab dishes or in animals. But eventually researchers hope to test these new drugs with children in clinical trials, so that these drugs may someday have a role in treating unfavorable histology Wilms tumors.

References
See all references for Wilms Tumor

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