



Wilms Tumor

What is cancer?

The body is made up of trillions of living cells. Normal body cells grow, divide to make new cells, and die in an orderly fashion. During the early years of a person's life, normal cells divide faster to allow the person to grow. After the person becomes an adult, most cells divide only to replace worn-out or dying cells or to repair injuries.

Cancer begins when cells in a part of the body start to grow out of control. There are many kinds of cancer, but they all start because of out-of-control growth of abnormal cells.

Cancer cell growth is different from normal cell growth. Instead of dying, cancer cells continue to grow and form new, abnormal cells. In most cases the cancer cells form a tumor. Cancer cells can also invade (grow into) other tissues, something that normal cells cannot do. Growing out of control and invading other tissues are what makes a cell a cancer cell.

Cells become cancer cells because of damage to DNA. DNA is in every cell and directs all its actions. In a normal cell, when DNA gets damaged the cell either repairs the damage or the cell dies. In cancer cells, the damaged DNA is not repaired, but the cell doesn't die like it should. Instead, this cell goes on making new cells that the body does not need. These new cells will all have the same damaged DNA as the first cell does.

People can inherit damaged DNA, but often the DNA damage is caused by mistakes that happen while the normal cell is reproducing or by something in our environment. In adults, sometimes the cause of the DNA damage is something obvious, like cigarette smoking. But often no clear cause is found.

Cancer cells often travel to other parts of the body, where they begin to grow and form new tumors that replace normal tissue. This process is called *metastasis*. It happens when the cancer cells get into the bloodstream or lymph vessels of our body.

Different types of cancer can behave very differently. They grow at different rates and respond to different treatments. That is why people with cancer need treatment that is aimed at their particular kind of cancer.

Not all tumors are cancerous. Tumors that aren't cancer are called *benign*. Benign tumors can cause problems – they can grow very large and press on healthy organs and tissues. But they cannot grow into (invade) other tissues. Because they can't invade, they also can't spread to other parts of the body (metastasize). These tumors are almost never life threatening.

What are the differences between cancers in adults and children?

The types of cancers that develop in children are often different from the types that develop in adults. Childhood cancers are often the result of DNA changes that take place 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 survive their 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. Being treated in these centers offers the advantage of 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, 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 our document *Children Diagnosed With Cancer: Dealing With Diagnosis*.

What is Wilms tumor?

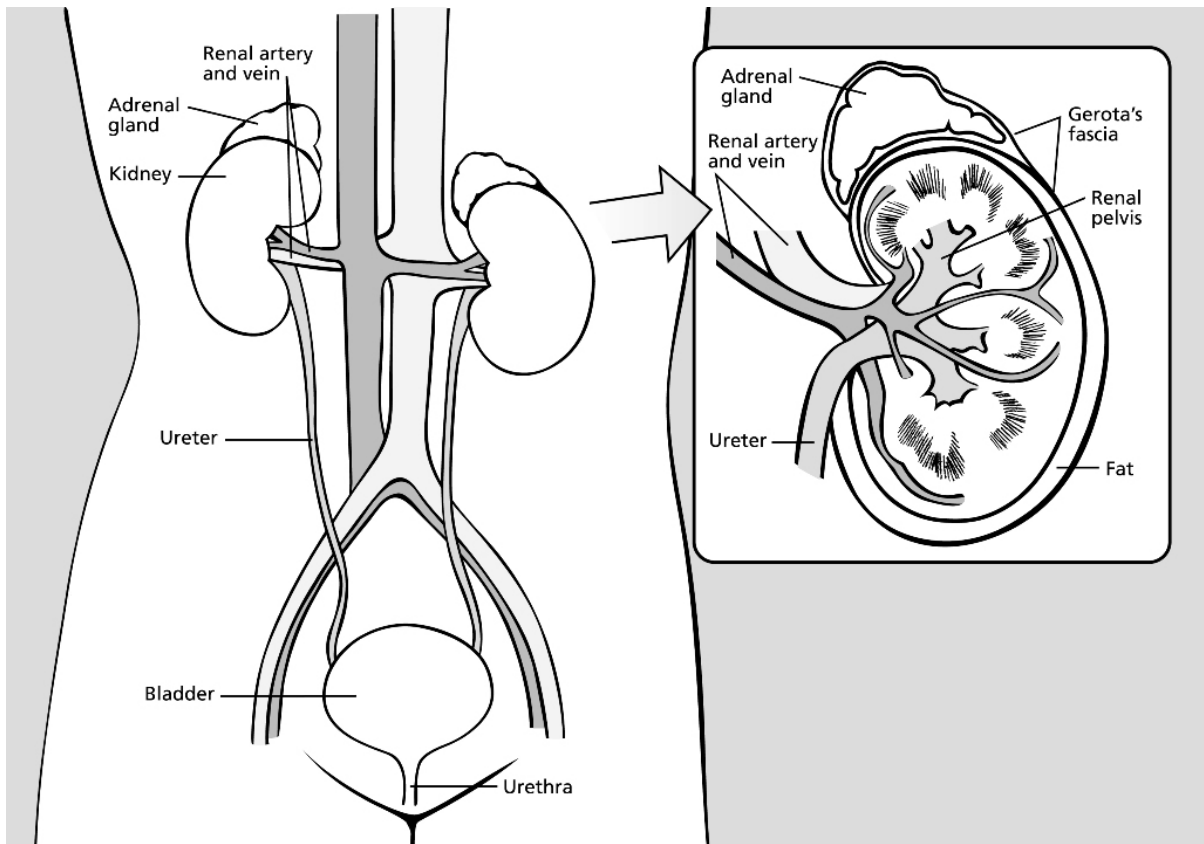
Wilms tumor (also called *Wilms' tumor* or *nephroblastoma*) is a type of cancer that starts in the kidneys. It is the most common type of kidney cancer in children. It is named after Max

Wilms, a German doctor who wrote one of the first medical articles about the disease in 1899.

About the kidneys

To understand Wilms tumor, it helps to know about the kidneys and what they do.

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



The kidneys' main job is to filter the blood and rid the body of excess water, salt, and waste products. The filtered products and extra water are changed into 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. They do this by making a hormone called *erythropoietin*, which tells the bone marrow to make more red blood cells.

Our kidneys are important, but we actually need less than one complete kidney to do all of its basic functions. Tens of thousands of people in the United States are living normal, healthy lives with just one kidney.

Wilms tumors

Wilms tumors are the most common cancers in children that start in the kidneys. 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 (cancer 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 tumors are found before they have spread (metastasized) to other organs.

Even though a doctor might think a child has a cancer such as Wilms tumor based on a physical exam or imaging tests, they cannot be certain until a sample of the tumor is looked at under a microscope.

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 cure for 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

About 9 of 10 kidney cancers in children are Wilms tumors, but in rare cases children may develop other types of kidney tumors.

Mesoblastic nephroma

These tumors usually appear in the first few months of life. Patients 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 is usually similar to 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 completely removed 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.

The rest of this document refers only to 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."

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 are 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

The risk of Wilms tumor is slightly higher 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 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):

- **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. Children with WAGR tend to get Wilms tumors at an earlier age and often have bilateral disease (tumors in both kidneys).

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 (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 also caused by a defect in chromosome 11.

Denys-Drash syndrome

This rare syndrome has been linked to changes (mutations) in the *WT1* gene. In this 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 do not develop normally, which in boys may cause them to 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.

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
- Frasier 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)

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 may 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 does not happen, the cells may 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 is the chemical in each of our cells that makes up our genes – the instructions for how our cells function.

Some genes control when our cells grow, divide into new cells, and die. Certain 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 other growth abnormalities, like some of those described in the section "What are the risk factors for Wilms tumor?" are sometimes found together with Wilms tumors.

Sometimes these gene changes are passed on from a parent to a child, but most Wilms tumors do not seem to be caused by inherited gene mutations. Instead, they seem to be the result of 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 *WTX*, which is found on the X chromosome. Another gene that is sometimes altered in Wilms tumor cells is known as *CTNNB1*. It's not clear exactly what causes these genes to be altered.

Because the genes described above are not altered in all cases of Wilms tumors, there must be changes in other genes 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 might cause these changes. Some gene changes may 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 remember that there is nothing these children or their parents could have done to prevent these cancers.

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) cannot 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 that a mother could avoid.

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.

Can Wilms tumor be found early?

Wilms tumors are usually found when they start to cause symptoms such as swelling in the abdomen (belly), but by this point they have often grown quite large. They can be found earlier in some children with tests such as an ultrasound of the abdomen (see the section "How are Wilms tumors diagnosed?"). But because Wilms tumors are so rare, it's not practical to do an ultrasound exam as a screening test (a test to look for disease in people with no signs or symptoms) in all children who are not at increased risk. There are no blood tests or other tests that are useful in screening otherwise healthy children for Wilms tumors.

On the other hand, screening for Wilms tumor is very important for children who have syndromes or birth defects known to be linked to this disease. For these children, most doctors recommend physical exams by a specialist and ultrasound exams on a regular basis (for example, about every 3 or 4 months at least until the age of 8) to find any kidney tumors when they are still small and have not yet spread to other organs.

Wilms tumor can also run in families, although this is rare. Talk to your doctor if you have any relatives who have had a Wilms tumor. If you do, the children in your family may need to have regular ultrasound exams. If a man or woman is known to have a *WT1* gene mutation, testing can be done to see if they have passed the mutation on to their children. (This can be done even before birth.)

Signs and symptoms of Wilms tumor

Wilms tumors can be hard to find early because they can often grow quite large without causing any symptoms. Children may look healthy and play normally.

Swelling or a hard mass in the abdomen (belly): This is often the first sign of a Wilms tumor. Parents may notice this while bathing or dressing the child. It feels firm and is often large enough to be felt on both sides of the belly. It's usually not painful, but it might cause belly pain in some cases.

Other possible symptoms: Some children with Wilms tumor may also have:

- Fever
- Nausea
- Loss of appetite
- Shortness of breath
- Constipation
- Blood in the urine

Wilms tumors can also sometimes cause high blood pressure. This does not usually cause symptoms on its own, but in rare cases it can get high enough to cause problems such as bleeding inside the eye or even a change in consciousness.

Many of the signs and symptoms above are more likely to be caused by something other than a kidney tumor. Still, if your child has any of these symptoms, check with your child's doctor so that the cause can be found and treated, if needed.

How are Wilms tumors diagnosed?

Wilms tumors are usually found when a child is brought to a doctor because of symptoms he or she is having. The doctor might suspect a child has a Wilms tumor based on a physical exam or other tests, but the diagnosis can only be made for certain once a sample of the tumor is removed and looked at under a microscope.

Medical history and physical exam

If your child has signs or symptoms that suggest he or she may have a kidney tumor, the doctor will want to get a complete medical history to learn more about the symptoms and how long they have been there. The doctor may also ask if there's a family history of cancer or birth defects, especially in the genitals or urinary system.

The doctor will examine your child to look for possible signs of a kidney tumor or other health problems. The main focus will likely be on the abdomen and on any increase in blood pressure, which is another possible sign of a kidney tumor. Blood and urine samples might also be collected at this time for testing (see "Lab tests" below).

Imaging tests

If the doctor thinks your child might have a kidney tumor, he or she will probably order one or more of the imaging tests below. These tests use sound waves, x-rays, magnetic fields, or radioactive substances to create pictures of the inside of the body. Imaging tests are done for a number of reasons, including:

- To help find out if there is a tumor in the kidney(s), and if so, if it is likely to be a Wilms tumor
- To learn how far the tumor may have spread, both within the kidney and to other parts of the body
- To help guide surgery or radiation therapy
- To look at the area after treatment to help determine if it has worked

For more details on the imaging tests discussed here, see our document *Imaging (Radiology) Tests*.

Ultrasound (sonogram)

This is often the first imaging test done if the doctor suspects your child has a Wilms tumor because it's easy to have, it does not use radiation, and it gives the doctor a good view of the kidneys and the other organs in the abdomen.

Ultrasound uses sound waves to create images of internal organs. For this test, your child lies on a table while a small wand called a *transducer* is placed on the skin (which is first lubricated with a gel) over the belly. It gives off sound waves and picks up the echoes as they bounce off the kidney. The echoes are converted by a computer into a black and white image on a screen.

The echo patterns made by most kidney tumors look different from those of normal kidney tissue. Different echo patterns also can help doctors tell some types of cancerous and non-cancerous kidney tumors apart from one another.

Ultrasound is also very useful when looking for tumor thrombus (tumor growing into the main veins coming out of the kidney). This helps in planning for surgery, if it is needed.

The test is not usually painful, but it might cause some discomfort if the transducer is pressed down hard on the abdomen.

Computed tomography (CT, CAT) scan

The CT scan is an x-ray test that produces detailed cross-sectional images of parts of your child's body, including organs such as the kidneys. This is one of the most useful tests to

look for a mass inside the kidney. It's also helpful in checking whether a cancer has grown into nearby veins or has spread to organs and tissues beyond the kidney, such as the lungs.

Instead of taking one picture, like a regular x-ray, a CT scanner takes many pictures as it rotates around your child while he or she lies on a table. A computer then combines these pictures into images showing slices of the part of the body being studied.

Before the scan, your child may be asked to drink a contrast solution and/or get an intravenous (IV) injection of a contrast dye that better outlines abnormal areas in the body. Your child may need an IV line for the dye. The contrast may cause some flushing (a feeling of warmth, especially in the face). Some people are allergic and get hives. Rarely, more serious reactions like trouble breathing or low blood pressure can occur. Be sure to tell the doctor if your child has any allergies or has ever had a reaction to any contrast material used for x-rays.

CT scans take longer than regular x-rays. A CT scanner has been described as a large donut, with a narrow table in the middle opening. Your child will need to lie still on the table while the scans are being done. During the test, the table slides in and out of the scanner. Younger children may be given medicine to help keep them calm or even asleep during the test to help make sure the pictures come out well.

Magnetic resonance imaging (MRI) scan

An MRI scan might be done if the doctor needs to see very detailed images of the kidney or nearby areas. For example, it might be done if there's a chance that a kidney tumor might have reached a major vein (the inferior vena cava) in the abdomen. An MRI scan might also be used to look for possible spread of cancer to the brain or spinal cord if doctors are concerned the cancer may have spread there.

Like CT scans, MRI scans provide detailed images of soft tissues in the body. But MRI scans use radio waves and strong magnets to create the images instead of x-rays and don't expose your child to radiation.

A contrast material called *gadolinium* may be injected into a vein before the scan to better see details. It usually does not cause allergic reactions, but it can cause other problems in children with kidney disease, so doctors are careful when they use it.

MRI scans take longer than CT scans – often up to an hour. Your child may have to lie inside a narrow tube, which is confining and can be distressing. Newer, more open MRI machines may help with this, but the test still requires staying still for long periods of time. The MRI machine also makes loud buzzing and clicking noises that your child may find disturbing. Younger children may be given medicine to help keep them calm or even asleep during the test.

Chest x-ray

Chest x-rays may be done to look for any spread of Wilms tumor to the lungs, as well as to have a baseline view of the lungs to compare with other x-rays that might be done in the future. If a CT scan of the chest is done, this test is not needed.

Bone scan

Bone scans can help show if cancer has spread to bones. Doctors don't usually order this test unless they think your child has a type of Wilms tumor that is likely to spread.

For this test, a small amount of low-level radioactive material is injected into a vein (intravenously, or IV). (The amount of radioactivity used is very low and will pass out of the body within a day or so.) The substance settles in areas of damaged bone throughout the skeleton over the course of a couple of hours. Your child then lies on a table for about 30 minutes while a special camera detects the radioactivity and creates a picture of the skeleton. Younger children may be given medicine to help keep them calm or even asleep during the test.

Areas of active bone changes will appear as hot spots on the skeleton – that is, they attract the radioactivity. These areas may suggest the presence of cancer, but other bone diseases can also cause the same pattern. To help tell these apart, other tests such as plain x-rays or MRI scans of the bone might be needed.

Lab tests

Lab tests might be done to check urine and blood samples if your child's doctor suspects a kidney problem. They may also be done after a diagnosis of Wilms tumor has been made.

A urine sample may be tested (urinalysis) to look for blood and other substances in the urine to see if there are problems with the kidneys. The urine may also be tested for substances called *catecholamines*. This is done to make sure your child doesn't have another kind of tumor called neuroblastoma. (Neuroblastomas often start in the adrenal gland, which lies just above the kidney.)

Blood tests are not used to find Wilms tumors, but they can sometimes show if a child has kidney problems. They can also be done to check a child's general health (especially before surgery) and to look for side effects during treatment such as chemotherapy. These may include tests to count the number of white blood cells, red blood cells, and blood platelets, and tests to measure certain chemicals and salts in the blood that give clues about how well the kidneys and liver are working.

Kidney biopsy/surgery

Most of the time, imaging tests can give doctors enough information to decide if a child probably has a Wilms tumor, and therefore if surgery should be done. But the actual diagnosis of Wilms tumor is made when a sample of the tumor is removed and looked at under a microscope. The cells in Wilms tumors have a distinct appearance when looked at this way. Doctors also look at the sample to determine the histology of the Wilms tumor (favorable or unfavorable), as was described in the section “What is Wilms tumor?”

In most cases, the sample is removed during surgery to treat the tumor (see the Surgery section). Sometimes if the doctors are less certain about the diagnosis or if they are not sure the tumor can be removed completely, a sample of the tumor may be taken during a biopsy as a separate procedure done before surgery. The biopsy may be done either as a type of surgery or using a long, hollow needle that’s inserted through the skin and into the tumor.

See *Testing Biopsy and Cytology Specimens for Cancer* to learn more about different types of biopsies, how the tissue is used in the lab for disease diagnosis, and what the results will tell you.

How is Wilms tumor staged?

The stage of a cancer describes how far it has spread. Your child’s treatment and prognosis (outlook) depend, to a large extent, on the cancer’s stage. Staging is based on the results of the physical exam and imaging tests (ultrasound, CT scans, etc.), which were described in “How is Wilms tumor diagnosed?”, as well as on the results of surgery to remove the tumor, if it has been done.

Children’s Oncology Group (COG) staging system

A staging system is a standard way for the cancer care team to sum up their findings of how extensive the tumor is. In the United States, the Children’s Oncology Group staging system is used most often to describe the extent of spread of Wilms tumors. This system describes Wilms tumor stages using Roman numerals I through V (1 through 5).

Stage I

The tumor was contained within one kidney and was completely removed by surgery. The tissue layer surrounding the kidney (the renal capsule) was not broken during surgery. The cancer had not grown into blood vessels in or next to the kidney. The tumor was not biopsied before surgery to remove it.

About 40% to 45% of all Wilms tumors are stage I.

Stage II

The tumor has grown beyond the kidney, either into nearby fatty tissue or into blood vessels in or near the kidney, but it was completely removed by surgery without any apparent cancer left behind. Lymph nodes do not contain cancer. The tumor was not biopsied before surgery.

About 20% of all Wilms tumors are stage II.

Stage III

This stage refers to Wilms tumors that may not have been completely removed. The cancer remaining after surgery is limited to the abdomen (belly). One or more of the following features may be present:

- The cancer has spread to lymph nodes (bean-sized collections of immune cells) in the abdomen or pelvis but not to more distant lymph nodes, such as those inside the chest.
- The cancer has invaded nearby vital structures so the surgeon could not remove it completely.
- Deposits of tumor (tumor implants) are found along the inner lining of the abdominal space.
- Cancer cells are found at the edge of the sample removed by surgery, indicating that some of the cancer still remains after surgery.
- Cancer cells “spilled” into the abdominal space before or during surgery.
- The tumor was removed in more than one piece – for example, the tumor was in the kidney and in the nearby adrenal gland, which was removed separately.
- A biopsy of the tumor was done before it was removed with surgery.

About 20% to 25% of all Wilms tumors are stage III.

Stage IV

The cancer has spread through the blood to organs away from the kidneys such as the lungs, liver, brain, or bone, or to lymph nodes far away from the kidneys.

About 10% of all Wilms tumors are stage IV.

Stage V

Tumors are found in both kidneys at diagnosis.

About 5% of all Wilms tumors are stage V.

Survival rates for Wilms tumor by stage and histology

Survival rates are often used by doctors as a standard way of discussing a person's prognosis (outlook). Some parents may want to know the survival statistics for children in similar situations, while others may not find the numbers helpful, or may even not want to know them. If you would rather not read about survival rates, please skip to the next section.

The 4-year survival rate refers to the percentage of children who live *at least* 4 years after their cancer is diagnosed. Of course, many children live much longer than 4 years (and many are cured).

In order to get 4-year survival rates, doctors have to look at children who were treated at least 4 years ago. Improvements in treatment since then may result in a better outlook for children now being diagnosed with Wilms tumors.

The survival rates below are based on the results of the National Wilms Tumor Studies, which included most of the children treated in the United States in the last few decades. The 2 most important factors in determining a child's outlook are the stage and histology of the tumor. (The histology refers to how the cancer cells look under the microscope – see “What is Wilms tumor?”) Some of these rates are based on only small numbers of cases, so they might not be accurate.

Survival rates are based on previous outcomes of children who had the disease, but they cannot predict what will happen in any particular child's case. Knowing the stage and histology of a child's Wilms tumor are important in estimating their outlook. But other factors may also affect a child's outlook, such as how well the tumor responds to treatment. Even when taking other factors into account, survival rates are only rough estimates. Your child's doctor can tell you if the numbers below apply, as he or she is familiar with your child's situation.

Wilms Tumor 4-year Survival Rates

Tumor Stage	Favorable Histology	Unfavorable Histology (Anaplastic Wilms Tumor)
I	99%	83%
II	98%	81%
III	94%	72%
IV	86%	38%
V	87%	55%

How is Wilms tumor treated?

This information represents the views of the doctors and nurses serving on the American Cancer Society's Cancer Information Database Editorial Board. These views are based on their interpretation of studies published in medical journals, as well as their own professional experience.

The treatment information in this document is not official policy of the Society and is not intended as medical advice to replace the expertise and judgment of your cancer care team. It is intended to help you and your family make informed decisions, together with your doctor.

Your doctor may have reasons for suggesting a treatment plan different from these general treatment options. Don't hesitate to ask him or her questions about your treatment options.

General comments about treatment

Overall, about 9 of 10 children with Wilms tumor are cured. A great deal of progress has been made in treating this disease with surgery, radiation therapy, and chemotherapy. Much of this progress in the United States has been because of the work of the National Wilms Tumor Study Group (now part of the Children's Oncology Group), which runs clinical trials of new treatments for children with Wilms tumor. Today, most children with this cancer are treated in a clinical trial to try to improve on what doctors believe is the best treatment. The goal of these studies is to find ways to cure as many children as possible while limiting side effects by giving as little treatment as needed.

Because Wilms tumors are rare, few doctors outside of those in children's cancer centers have much experience in treating them. Most doctors recommend a team approach that includes the child's pediatrician as well as specialists at a child's cancer center. For Wilms tumors, the doctors on this team often include:

- A pediatric surgeon or pediatric urologist (doctor who treats urinary system problems in children [and genital problems in boys])
- A pediatric oncologist (doctor who uses chemotherapy and other medicines to treat childhood cancers)
- A pediatric radiation oncologist (doctor who uses radiation therapy to treat cancer in children)

Many other specialists may be involved in your child's care as well, including physician assistants, nurse practitioners, nurses, psychologists, social workers, rehabilitation specialists, and other health professionals. For more information, see our document *Children Diagnosed With Cancer: Understanding the Health Care System*.

After your child's tumor is found and its stage and histology are determined, the cancer care team will discuss treatment options with you. It's important to discuss all of the options as well as their possible side effects with your child's doctors so you can make an informed

decision. (For a list of some questions to ask, see the section “What should you ask your child’s doctor about Wilms tumor?”)

If time permits, it can often be helpful to get a second opinion if you have questions about the recommended plan (or if you just want to confirm that it’s the best option). This can provide you with more information and help you feel more confident about the treatment plan you choose.

The main types of treatment that can be used for Wilms tumor are:

- Surgery
- Chemotherapy
- Radiation therapy

Most children will get more than one type of treatment.

In the United States, surgery is the first treatment for most Wilms tumors. In Europe, doctors prefer to give a short course of chemotherapy before the surgery. There seems to be no difference in the results from these 2 approaches.

The first goal of treatment is to remove the primary (main) tumor even if the cancer has spread to distant parts of the body. Sometimes the tumor may be hard to remove. It may be very large, it may have spread into nearby blood vessels or other vital structures, or it may be in both kidneys. For these children, doctors might use chemotherapy, radiation therapy, or a combination of the 2 to try to shrink the tumor(s) before surgery.

If any cancer cells remain after surgery, radiation therapy or more surgery may be needed.

The next few sections describe the types of treatments used for Wilms tumors. This is followed by a description of the most common approaches used for these tumors based on the tumor stage (extent) and histology (appearance under a microscope).

Please see the “Additional resources for Wilms tumor” section for other, more specific materials on the different types of cancer treatments and their side effects.

Surgery for Wilms tumors

Surgery is the main treatment for nearly all children with Wilms tumor. It should be done by a surgeon who specializes in operating on children and has experience in treating these cancers.

Removing the tumor

The main goal of surgery is to try to remove the entire Wilms tumor in one piece to prevent the possible spread of cancer cells within the abdomen (belly). Surgeons who operate on

these tumors are careful to limit the chance of this type of cancer spread whenever possible. If the surgeon finds (either with imaging tests done before surgery, or when starting the operation) that the entire tumor can't be removed safely, other treatments may be used first. If these treatments shrink the tumor enough, surgery can then be done more safely.

Depending on the situation, different operations may be used.

Radical nephrectomy: This is the most common surgery for a Wilms tumor that's only in one kidney, as it provides the best chance of making sure all of the tumor is removed. During this operation, the surgeon makes an incision (cut – usually down the middle of the abdomen) and removes the cancer along with the whole kidney, the fatty tissue around the kidney, the ureter (tube that carries urine from the kidney to the bladder), and the attached adrenal gland (a hormone-making gland that sits on top of the kidney). Most children do very well with only one kidney.

Partial nephrectomy (nephron-sparing surgery): In the small number of children who have Wilms tumors in both kidneys, the surgeon will try to save some normal kidney tissue, if possible. The surgeon may remove the kidney containing the most tumor with a radical nephrectomy. In the other kidney the surgeon may do a partial nephrectomy, removing just the tumor and a margin of normal kidney around it. Another option may be to do partial nephrectomies on both kidneys.

Sometimes, both kidneys may need to be removed completely. The child will then need dialysis several times a week. In this procedure, a machine does the job of the kidneys by filtering waste products out of the blood. Once the child is healthy enough, a kidney transplant may be an option if a donor kidney becomes available.

Assessing the extent of the disease (surgical exploration)

Another main goal of surgery (radical or partial nephrectomy) is to determine the extent of the cancer and whether or not it can all be removed. Lymph nodes near the kidney will be removed during surgery to look for cancer cells in them. (Lymph nodes are bean-sized collections of immune cells to which cancer often spreads.) Lymph node removal is known as a *regional lymphadenectomy*.

The other kidney and nearby organs such as the liver may also be looked at closely, and any suspicious areas biopsied (samples taken to be checked for cancer under a microscope).

Knowing if a Wilms tumor has spread to the lymph nodes, the other kidney, or other nearby organs is important in determining its stage and further treatment options.

Placing a central venous access device (port)

Often, if the child is going to get chemotherapy, a surgeon will insert a small plastic tube called a catheter into a large blood vessel – usually under the collar bone. This tube may be called a *venous access device*, *central venous catheter*, or just a *port*. The tube might be put

in during the main surgery to remove the tumor, or as a separate operation (especially if chemo is going to be given before the main surgery).

The end of the tube will be just under the skin or sticking out of the chest area or upper arm. This can be left in place for several months to take blood samples for tests and to give intravenous (IV) chemotherapy drugs and blood transfusions, without having to use a needle in the arm each time. Members of the cancer care team will teach you how to care for your child's venous access device to reduce the risk of problems, such as clotting and infections.

Possible risks and side effects of surgery

Surgery to remove a Wilms tumor is a serious operation, and surgeons are very careful to try to limit any problems either during or after surgery. Complications during surgery, such as bleeding, injuries to major blood vessels or other organs, or reactions to anesthesia, are rare, but they can happen.

Almost all children will have some pain for a while after the operation, although this can usually be helped with medicines if needed. Other problems after surgery are not common but can include internal bleeding, infections, or problems with food moving through the intestines.

If there are tumors in both kidneys, another concern is the loss of kidney function. In these cases, doctors must balance between making sure the tumors are removed completely and removing only as much of the kidney(s) as is needed. Children who have all or parts of both kidneys removed may need dialysis, and may eventually need a kidney transplant.

For more information on surgery as a treatment for cancer, see our document *A Guide to Cancer Surgery*.

Chemotherapy for Wilms tumors

Chemotherapy (chemo) uses anti-cancer drugs that are given into a vein or by mouth (in pill form). These drugs enter the blood and reach all areas of the body, which makes this treatment useful for cancer that has spread or might have spread beyond the kidney.

Most children with Wilms tumors will get chemotherapy at some point during their treatment. In the United States, chemo is usually given after surgery. Sometimes it may be needed before surgery to shrink a tumor to make the operation possible. In Europe, chemo is given before surgery and continued afterward. In both cases, the type and amount of chemo depend on the stage and histology of the cancer.

A combination of chemo drugs is used to treat children with Wilms tumors. The chemo drugs used most often are actinomycin D (dactinomycin) and vincristine. For tumors at more advanced stages, those with unfavorable histology, or tumors that recur (come back) after treatment, other drugs such as doxorubicin (Adriamycin), cyclophosphamide, etoposide, irinotecan, and/or carboplatin may also be used.

These drugs are injected into a vein or into a venous access device. Different drugs, doses, and lengths of treatment are used, depending on the type and stage of the Wilms tumor and the child's age. Most often, the drugs are given once a week for at least several months. They are usually given by a nurse in the doctor's office or in the outpatient section of the hospital. In some cases, children with Wilms tumors stay in the hospital while they are getting chemo, but usually this is not necessary.

Possible side effects of chemotherapy

Chemo drugs attack cells that are dividing quickly, which is why they often work against cancer cells. But other cells in the body, such as those in the bone marrow (where new blood cells are made), the lining of the mouth and intestines, and the hair follicles, also divide quickly. These cells are also likely to be affected by chemo, which can lead to side effects.

The side effects of chemo depend on the type of drugs, the amount taken, and the length of treatment. Possible short-term side effects can include:

- Hair loss
- Mouth sores
- Loss of appetite
- Nausea and vomiting
- Diarrhea or constipation
- Increased chance of infections (from having too few white blood cells)
- Easy bruising or bleeding (from having too few blood platelets)
- Fatigue or extreme tiredness (from having too few red blood cells)

Your child's doctor and treating team will watch closely for any side effects that develop. There are often ways to lessen these side effects. For example, drugs can be given to help prevent or reduce nausea and vomiting. Be sure to ask your child's doctor or nurse about medicines to help reduce side effects, and let him or her know if your child has side effects so they can be managed.

Along with the effects listed above, some drugs can have specific side effects. For example:

- Vincristine can damage nerves. Some patients may have tingling, numbness, weakness, or pain, particularly in the hands and feet.
- Doxorubicin may cause heart damage. The risk of this happening goes up as the total amount of the drug given goes up. Doctors try to limit this risk as much as possible by not giving more than the recommended doses and by checking the heart with a test called an *echocardiogram* (an ultrasound of the heart) during treatment.

- Cyclophosphamide can damage the bladder, which can cause blood in the urine. The risk of this can be lowered by giving the drug with plenty of fluids and with a drug called *mesna*, which helps protect the bladder.

Lab tests to check for chemo side effects

Before each chemotherapy session, your child's doctor will check blood test results to see how well the liver, kidneys, and bone marrow are working. If there are problems, the chemo might need to be delayed or the doses reduced. You can learn more about these tests and what they mean in *Understanding Your Lab Test Results*.

Long-term side effects of chemo

Possible long-term effects of treatment are one of the major challenges children might face after cancer treatment.

For example, if your child is given doxorubicin (Adriamycin), there is a chance it could damage the heart. Your child's doctor will carefully watch the doses used and will check your child's heart function with imaging tests.

Some chemo drugs can increase the risk of developing a second type of cancer (such as leukemia) years after the Wilms tumor is cured. But this small increase in risk has to be weighed against the importance of chemotherapy in treating Wilms tumor. (See *Second Cancers Caused by Cancer Treatment* to find out more about this.)

Some drugs might also affect fertility (the ability to have children) years later.

See the section "What happens after treatment for Wilms tumor?" for more on the possible long-term effects of treatment.

For more information on chemotherapy in general, see the "Chemotherapy" section of our website, or our document *A Guide to Chemotherapy*.

Radiation therapy for Wilms tumor

Radiation therapy uses high-energy rays or particles to kill cancer cells. It is usually part of treatment only for more advanced Wilms tumors (stages III, IV, and V) and for some earlier stage tumors with unfavorable histology.

The type of radiation used for Wilms tumors, known as *external beam radiation therapy*, focuses radiation from outside the body on the cancer. It is much like getting an x-ray, although the dose of radiation is much higher. The total dose of radiation is divided into fractions, usually given 5 days a week for a couple of weeks.

Before treatments start, the radiation team takes careful measurements with imaging tests such as CT or MRI scans to determine the correct angles for aiming the radiation beams and the proper dose of radiation. Your child may be fitted with a plastic mold that looks like a

body cast to keep him or her in the same position during each treatment so that the radiation can be aimed more accurately.

For each treatment session, your child lies on a special table while a machine delivers the radiation from a precise angle. Each session lasts about 15 to 30 minutes, with most of the time being spent making sure the radiation is aimed correctly. The actual treatment time each day is much shorter. The treatment is not painful, but some younger children may be given medicine to make them drowsy before each treatment to help make sure they stay still.

Newer radiation techniques

Some newer techniques can help doctors aim the treatment at the tumor more accurately while reducing the radiation exposure to nearby healthy tissues. These techniques may help increase the success rate and reduce side effects.

Three-dimensional conformal radiation therapy (3D-CRT): 3D-CRT uses the results of imaging tests such as MRI and special computers to precisely map the location of the tumor. Several radiation beams are then shaped and aimed at the tumor from different directions. Each beam alone is fairly weak, which makes it less likely to damage normal body tissues, but the beams converge at the tumor to give a higher dose of radiation there.

Intensity modulated radiation therapy (IMRT): IMRT is an advanced form of 3D therapy. Along with shaping the beams and aiming them at the tumor from several angles, the intensity (strength) of the beams can be adjusted to limit the dose reaching the nearby normal tissues. This lets the doctor deliver a higher dose to the tumor. Many major hospitals and cancer centers now use IMRT.

Possible side effects of radiation therapy

Radiation is often an important part of treatment, but young children's bodies are very sensitive to it, so doctors try to use as little as possible to help avoid or limit any problems. Radiation therapy can cause both short-term and long-term side effects, which depend on the dose of radiation and where it's aimed.

Possible short-term effects:

- Effects on skin areas that receive radiation can range from mild sunburn-like changes and hair loss to more severe skin reactions.
- Radiation to the abdomen (belly) can cause nausea or diarrhea.
- Radiation therapy can make a child tired, especially toward the end of treatment.

Possible long-term effects:

- Radiation can slow the growth of normal body tissues (such as bones) that get radiation, especially in younger children. In the past this led to problems such as short bones or a curving of the spine, but this is less likely with the lower doses of radiation used today.
- Radiation that reaches the chest area can affect the heart and lungs. This does not usually cause problems right away, but in some children it might lead to heart or lung problems as they get older.
- In girls, radiation may damage the ovaries. This might lead to abnormal menstrual cycles or problems getting pregnant or having children later on.
- Radiation slightly increases the risk of developing a second cancer in the area, usually many years after it is given. This doesn't happen often with Wilms tumor because the amount of radiation used is low.

See the section “What happens after treatment for Wilms tumor?” for more on the possible long-term effects of treatment.

For more detailed information on radiation therapy, see the “Radiation” section of our website, or our document *Understanding Radiation Therapy: A Guide for Patients and Families*.

Clinical trials for Wilms tumor

You may have had to make a lot of decisions since you've been told your child has cancer. One of the most important decisions you will make is deciding which treatment is best. You may have heard about clinical trials being done for this type of cancer. Or maybe someone on your health care team has mentioned a clinical trial to you.

Clinical trials are carefully controlled research studies that are done with patients who volunteer for them. These studies are done to get a closer look at promising new treatments or procedures.

If you would like to find out more about clinical trials your child may be eligible for, you should start by asking your doctor if your clinic or hospital conducts clinical trials. Children's cancer centers often conduct many clinical trials at any one time, and most children treated at these centers take part in a clinical trial as part of their treatment.

You can also call our clinical trials matching service for a list of clinical trials that may meet your child's needs. You can reach this service at 1-800-303-5691 or on our website at www.cancer.org/clinicaltrials. You can also get a list of current clinical trials by calling the National Cancer Institute Cancer Information Service toll free at 1-800-4-CANCER (1-800-422-6237) or by visiting the NCI clinical trials website www.cancer.gov/clinicaltrials.

Your child will have to meet certain requirements to take part in any clinical trial. If your child does qualify for a clinical trial, you will have to decide whether or not to enter (enroll)

the child into it. Older children, who can understand more, usually must also agree to take part in the clinical trial before the parents' consent is accepted.

Clinical trials are one way to get state-of-the-art cancer care for your child. Sometimes they may be the only way to get access to some newer treatments. They are also the only way for doctors to learn better methods to treat cancer. Still, they might not be right for every child.

You can get a lot more information on clinical trials in our document *Clinical Trials: What You Need to Know*. You can read it on our website or call us at (1-800-227-2345) to have it sent to you.

Complementary and alternative therapies for Wilms tumor

When your child has cancer you may hear about ways to treat his or her cancer or relieve symptoms that your doctor hasn't mentioned. Everyone from friends and family to Internet groups and websites may offer ideas for what might help. These methods can include vitamins, herbs, and special diets, or other methods such as acupuncture or massage, to name a few.

What exactly are complementary and alternative therapies?

Not everyone uses these terms the same way, and they are used to refer to many different methods, so it can be confusing. We use *complementary* to refer to treatments that are used *along with* your regular medical care. *Alternative* treatments are used *instead of* a doctor's medical treatment.

Complementary methods: Most complementary treatment methods are not offered as cures for cancer. Mainly, they are used to help the person with cancer feel better. Some methods that are used along with regular treatment are: art therapy or play therapy to reduce stress; acupuncture to help relieve pain; or peppermint tea to relieve nausea. Some complementary methods are known to help, while others have not been tested. Some have been proven not to be helpful, and a few have even been found harmful.

Alternative treatments: Alternative treatments may be offered as cancer cures. These treatments have not been proven safe and effective in clinical trials. Some of these methods may pose danger, or have life-threatening side effects. But the biggest danger in most cases is that your child may lose the chance to be helped by standard medical treatment. Delays or interruptions in medical treatments may give the cancer more time to grow and make it less likely that treatment will help.

Finding out more

It is easy to see why parents who have children with cancer think about alternative methods. You want to do all you can to help fight the cancer, and the idea of a treatment with few or no side effects sounds great. Sometimes medical treatments like chemotherapy can be hard to

take, or they may no longer be working. But the truth is that most of these alternative methods have not been tested and proven to work in treating cancer.

As you consider your child's options, here are 3 important steps you can take:

- Look for “red flags” that suggest fraud. Does the method promise to cure all or most cancers? Are you told not to use regular medical treatments? Is the treatment a “secret” that requires you to take your child to certain providers or to another country?
- Talk to your child's doctor or nurse about any method you are thinking about.
- Contact us at 1-800-227-2345 to learn more about complementary and alternative methods in general and to find out about the specific methods you are looking at. You can also read about them in the “Complementary and Alternative Medicine” section of our website.

The choice is yours

You always have a say in how your child is treated. If you want to use a non-standard treatment, learn all you can about the method and talk to your child's doctor about it. With good information and the support of your child's health care team, you may be able to safely use the methods that can help your child while avoiding those that could be harmful.

Treatment by type and stage of Wilms tumor

In the United States, most children with Wilms tumors are treated in clinical trials developed by the Children's Oncology Group. The goal of these studies is to cure as many children as possible while limiting side effects by giving as little treatment as is necessary. This is done by comparing the current best treatment with one the doctors think might be better. Because of this, treatment may differ slightly from what is described here in some cases.

Treatment for Wilms tumor is based mainly on the stage of the cancer and whether its histology (appearance under the microscope) is favorable or unfavorable. In the United States, doctors prefer to use surgery as the first treatment in most cases, and then give chemotherapy (and possibly radiation therapy) afterward. In Europe, doctors prefer to start the chemotherapy before surgery. The results seem to be about the same.

Most often, the stage and histology of the cancer are actually determined when surgery is done to remove the cancer, because the true extent of the tumor often can't be determined by imaging tests alone. The findings from surgery are then used to guide further treatment. But sometimes it's clear that the cancer has already spread beyond the kidney even before surgery is done, based on imaging tests. This can affect the order in which treatments are given, as well as the extent of surgery.

Stage I

These tumors are only in the kidney. Surgery has completely removed the tumor along with the entire kidney, nearby structures, and some nearby lymph nodes.

Favorable histology: Children younger than 2 years with small tumors (weighing less than 550 grams) may not need further treatment, such as chemo. But they need to be watched closely because the chance the cancer will come back is slightly higher than if they had chemo. If the cancer does come back, the chemo drugs actinomycin D (dactinomycin) and vincristine (and possibly more surgery) are very likely to be effective at this point.

For children older than 2 and for those of any age who have larger tumors, surgery is usually followed by chemo with actinomycin D and vincristine. If the tumor cells have certain chromosome changes, the drug doxorubicin (Adriamycin) may be given as well. The chemo is given for several months.

Unfavorable histology: For children of any age who have tumors with unfavorable histology, surgery is usually followed by radiation therapy to the area of the tumor, along with chemo with actinomycin D, vincristine, and doxorubicin (Adriamycin) for several months.

Stage II

These tumors have grown outside the kidney into nearby tissues, but surgery removed all visible signs of cancer.

Favorable histology: After surgery, standard treatment is chemo with actinomycin D and vincristine. If the tumor cells have certain chromosome changes, the drug doxorubicin (Adriamycin) may be given as well. The chemo is given for several months.

Unfavorable histology with focal (only a little) anaplasia: When the child recovers from surgery, radiation therapy is given over several weeks. When this is finished, chemo (doxorubicin, actinomycin D, and vincristine) is given for about 6 months.

Unfavorable histology with diffuse (widespread) anaplasia: After surgery, these children get radiation over several weeks. This is followed by a more intense type of chemo using the drugs vincristine, doxorubicin, etoposide, cyclophosphamide, and carboplatin, along with mesna (a drug that protects the bladder from the effects of cyclophosphamide), which is given for about 6 months.

Stage III

These tumors were not removed completely with surgery because of their size or location or for other reasons. In some cases, surgery to remove the tumor may be postponed until other treatments are able to shrink the tumor first (see below).

Favorable histology: Treatment is usually surgery if it can be done, followed by radiation therapy over several days. This is followed by chemo with 3 drugs (actinomycin D, vincristine, and doxorubicin). If the tumor cells have certain chromosome changes, the drugs cyclophosphamide and etoposide may be given as well. Chemo is given for about 6 months.

Unfavorable histology with focal (only a little) anaplasia: Treatment starts with surgery if it can be done, followed by radiation therapy over several weeks. This is followed by chemo, usually with 3 drugs (actinomycin D, vincristine, and doxorubicin) for about 6 months.

Unfavorable histology with diffuse (widespread) anaplasia: Treatment starts with surgery if it can be done, followed by radiation therapy over several weeks. This is followed by chemo, usually with the drugs vincristine, doxorubicin, etoposide, cyclophosphamide, and carboplatin, along with mesna (a drug that protects the bladder from the effects of cyclophosphamide). Chemo lasts about 6 months.

In some instances the tumor may be very large or may have grown into nearby blood vessels or other structures so that it cannot be removed safely. In these children, a small biopsy sample is taken from the tumor to be sure that it's a Wilms tumor and to determine its histology. Then chemo is started. Usually the tumor will shrink enough within several weeks so that surgery can be done. If not, then radiation therapy might be given as well. Chemo will be started again after surgery. If radiation was not given before surgery, it's given after surgery.

Stage IV

These tumors have already spread to distant parts of the body at the time of diagnosis. As with stage III tumors, surgery to remove the tumor might be the first treatment, but it might need to be delayed until other treatments can shrink the tumor (see below).

Favorable histology: Surgery to remove the tumor is the first treatment if it can be done, followed by radiation therapy. The entire abdomen will be treated if there is still some cancer left after surgery. If the cancer has spread to the lungs, low doses of radiation might also be given to that area. This is followed by chemo, usually with 3 drugs (actinomycin D, vincristine, and doxorubicin) for about 6 months. If the tumor cells have certain chromosome changes, the drugs cyclophosphamide and etoposide may be given as well.

Unfavorable histology: Treatment might start with surgery if it can be done, followed by radiation therapy. The entire abdomen will be treated if there is still some cancer left after surgery. Low doses of radiation will also be given to both lungs if there is spread to the lungs. This is followed by chemo with the drugs vincristine, doxorubicin, etoposide, cyclophosphamide, and carboplatin, along with mesna given for about 6 months. If the tumor cells have diffuse (widespread) anaplasia, some doctors might try the chemo drugs irinotecan and vincristine first instead (although this is not yet a commonly used treatment). The treatment would then be adjusted based on if the tumor shrinks in response to these drugs.

If the tumor is too large or has grown too much to be treated safely with surgery first, a small biopsy sample may be taken from the tumor to be sure that it's a Wilms tumor and to determine its histology. Chemo and/or radiation therapy may then be used to shrink the tumor. Surgery may be an option at this point. This would be followed by more chemo and radiation therapy if it wasn't given already.

For stage IV cancers that have spread to the liver, surgery may be an option to remove any liver tumors that still remain after chemo and radiation therapy.

Stage V

Treatment for children with tumors in both kidneys is unique for each child, although it typically includes surgery, chemo, and radiation therapy at some point.

Biopsies (tissue samples) of tumors in both kidneys and of nearby lymph nodes may be taken first, although not all doctors feel this is needed because when both kidneys have tumors, the chance that they are Wilms tumors is very high.

Chemo is typically given first to try to shrink the tumors. The drugs used will depend on the extent and histology (if known) of the tumors. After about 6 weeks of chemo, surgery (partial nephrectomy) may be done to remove the tumors if enough normal kidney tissue can be left behind. If the tumors haven't shrunk enough, treatment may include more chemo or radiation therapy for about another 6 weeks. Surgery (either partial or radical nephrectomy) may then be done. This is followed by more chemo, possibly along with radiation therapy if it hasn't been given already.

If not enough functioning kidney tissue is left after surgery, a child may need to get dialysis, a procedure where a special machine filters waste products out of the blood several times a week. If there is no evidence of any cancer after a year or two, a donor kidney transplant may be done.

Recurrent Wilms tumor

The prognosis and treatment for children with Wilms tumor that recurs (comes back after treatment) depends on their prior treatment, the cancer's histology (favorable or unfavorable), and where it recurs. The outlook is generally better for recurrent Wilms tumors with the following features:

- Favorable histology
- Initial stage of I or II
- Initial chemo with vincristine and actinomycin D only
- No previous radiation therapy

The usual treatment for these children is surgery to remove the recurrent cancer (if possible), radiation therapy (if not already given to the area), and chemo, often with different drugs than those used during first treatment.

Recurrent Wilms tumors that do not have the features listed above are much harder to treat. These children are usually treated with aggressive chemo, such as the ICE regimen (ifosfamide, carboplatin, and etoposide) or others being studied in clinical trials. Very high-dose chemo followed by a stem cell transplant (sometimes called a *bone marrow transplant*) might also be an option in this situation. (See *Stem Cell Transplant (Peripheral Blood, Bone Marrow, and Cord Blood Transplants* for more on this treatment option.)

More treatment information for Wilms tumor

For more details on treatment options—including some that may not be addressed in this document—the National Cancer Institute (NCI) and the Children’s Oncology Group (COG) are good sources of information.

The NCI provides treatment guidelines via its telephone information center (1-800-4-CANCER) and its website (www.cancer.gov). Detailed guidelines intended for use by cancer care professionals are also available on www.cancer.gov.

The COG is the world’s largest organization devoted to childhood cancer research. The COG website, www.childrensoncologygroup.org, provides key information to help support children and their families from the time of diagnosis, through treatment, and beyond.

What should you ask your child’s doctor about Wilms tumor?

It is important to have frank, open discussions with your child’s cancer care team. You should ask any questions that are on your mind, no matter how small they may seem. Below are some questions to consider:

- What kind of kidney cancer does my child have? Is it a Wilms tumor?
- Is the histology of the tumor favorable or unfavorable?
- What is the stage of my child’s cancer, and what does that mean?
- Do we need to have other tests done before we can decide on treatment?
- How much experience do you have treating this type of cancer?
- What other doctors will we need to see?
- What are our treatment options?

- Are there any clinical trials we might want to consider?
- What do you recommend and why?
- What are the risks and side effects of the suggested treatments?
- Which side effects start shortly after treatment and which ones might develop later on?
- How might treatment affect my child's ability to grow and develop?
- Could treatment affect my child's ability to have children later on?
- Will my child have a higher long-term risk kidney problems or of other cancers?
- What should we do to be ready for treatment?
- How long will treatment last? What will it involve? Where will it be done?
- How will treatment affect our daily activities (school, work, etc.)?
- Based on what you've learned about my child's cancer, what is the outlook for cure?
- What would we do if the treatment doesn't work or if the cancer comes back?
- What type of follow-up will my child need after treatment?

Along with these sample questions, be sure to write down any others you might have. For instance, you might want to ask about getting a second opinion, or you may want more information about recovery times so you can plan your child's school and activity schedule. You might also want to ask about nearby or online support groups, where you may be able to get in touch with other families who have been through this.

What happens after treatment for Wilms tumor?

After treatment for Wilms tumor, the main concerns for most families are the short- and long-term effects of the tumor and its treatment, and concerns about the tumor still being present or coming back.

It's certainly normal to want to put the tumor and its treatment behind you, and get back to a life that doesn't revolve around cancer. But it's important to realize that follow-up care is a central part of treatment that offers your child the best chance for long-term recovery.

Follow-up exams and tests

Your child's health care team will set up a follow-up schedule, which will include physical exams and imaging tests (such as chest x-rays, ultrasounds, and CT scans) to look for the growth or return of the tumor, or any problems related to treatment. Since most children have had a kidney removed, blood and urine tests will be done to check how well the remaining kidney is working. If your child received doxorubicin (Adriamycin) during chemotherapy, the doctor may also order tests to check the function of your child's heart.

The recommended schedule for follow-up exams and tests depends on the initial stage and histology (favorable or unfavorable) of the cancer, the type of treatment, and any problems that the child may have had during treatment. Doctor visits and tests will be more frequent at first (about every 6 to 12 weeks for the first couple of years), but the time between visits may be extended as time goes on.

During this time, it's important to report any new symptoms to your child's doctor right away, so that the cause can be found and treated, if needed. Your child's doctor can give you an idea of what to watch for.

If the tumor does come back, or if it doesn't respond to treatment, your child's doctors will discuss the treatment options with you.

Children with bilateral Wilms tumor or Denys-Drash syndrome can develop kidney failure. These children will also need routine tests at least once a year to look for possible early signs of this (including urine tests, blood pressure checks, and blood tests of kidney function).

Late and long-term effects of treatment

Because of major advances in treatment, most children treated for Wilms tumor are now surviving into adulthood. Doctors have learned that treatment can affect children's health later in life, so watching for health effects as they get older has become more of a concern in recent years.

Just as the treatment of childhood cancer requires a very specialized approach, so does the care and follow-up after treatment. The earlier any problems can be recognized, the more likely it is they can be treated effectively.

Young people treated for Wilms tumor are at risk, to some degree, for several possible late effects of their cancer treatment. It's important to discuss what these possible effects might be with your child's medical team so you know what to watch for and report to the doctor.

The risk of late effects depends on a number of factors, such as the specific treatments the child has, the doses of treatment, and the age of the child when being treated. These late effects may include:

- Reduced kidney function

- Heart or lung problems after receiving certain chemotherapy drugs or radiation therapy to these parts of the body
- Slowed or delayed growth and development
- Changes in sexual development and ability to have children (especially in girls)
- Increased risk of second cancers later in life (rare)

To help increase awareness of late effects and improve follow-up care of childhood cancer survivors throughout their lives, the Children's Oncology Group (COG) has developed long-term follow-up guidelines for survivors of childhood cancers. These guidelines can help you know what to watch for, what type of screening tests should be done to look for problems, and how late effects may be treated.

Ask your child's health care team about possible long-term complications and make sure there's a plan in place to watch for these problems and treat them, if needed. To learn more, ask your child's doctors about the COG survivor guidelines. You can also download them for free at the COG website: www.survivorshipguidelines.org. The guidelines are written for health care professionals. Patient versions of some of the guidelines are available (as "Health Links") on the site as well, but we urge you to review them with a doctor.

For more about some of the possible long-term effects of cancer treatment, see our document *Children Diagnosed With Cancer: Late Effects of Cancer Treatment*.

Emotional and social issues for Wilms tumor survivors and their families

Most children with Wilms tumors are very young at the time of diagnosis. Still, some children may have emotional or psychological issues that need to be addressed during and after treatment. Depending on their age, they may also have some problems with normal functioning and school work. These can often be overcome with support and encouragement. Doctors and other members of the health care team can also often recommend special support programs and services to help children after treatment.

Parents and other family members can also be affected, both emotionally and in other ways. Some common family concerns include financial stresses, traveling to and staying near the cancer center, the possible loss of a job, and the need for home schooling.

Centers that treat many patients with Wilms tumors may have programs to introduce new patients and their families to others who have finished their treatment. This can give parents an idea of what to expect during and after treatment, which is very important. Seeing another patient with Wilms tumor doing well is often helpful.

Support groups for families of children with cancer can also be helpful. If you need help finding such a group, call your American Cancer Society at 1-800-227-2345 and we can put you in touch with a group or resource that may work for you.

Keeping good medical records

As much as you may want to put the experience behind you once treatment is completed, it's very important to keep good records of your child's medical care during this time. Gathering these details soon after treatment may be easier than trying to get them at some point in the future. There is certain information that your child's doctors should have, even into adulthood, including:

- A copy of the pathology report(s) from any biopsies or surgeries.
- If your child had surgery, a copy of the operative report(s).
- If your child stayed in the hospital, a copy of the discharge summaries doctors prepare when patients are sent home.
- If chemotherapy was given, a list of the final doses of each drug your child received. (Certain drugs may have specific long-term side effects. If you can get a list of these from the pediatric oncologist, it might help any new doctors your child has in the future.)
- If radiation therapy was given, a summary of the type and dose of radiation and when and where it was given.

It's also very important to keep your health insurance. Follow-up tests and doctor visits cost a lot, and even though no one wants to think of the tumor coming back, this could happen.

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 intensive 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 intensive 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 patients 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 in clinical trials, so that they may someday have a role in treating children with unfavorable histology Wilms tumors.

Additional resources for Wilms tumor

More information from your American Cancer Society

Here is more information you might find helpful. You also can order free copies of our documents from our toll-free number, 1-800-227-2345, or read them on our website, www.cancer.org.

Children with cancer

[Children Diagnosed With Cancer: Dealing with Diagnosis](#) (also in Spanish)

[Pediatric Cancer Centers](#) (also in Spanish)

[Children Diagnosed With Cancer: Understanding the Health Care System](#) (also in Spanish)

[Children Diagnosed With Cancer: Financial and Insurance Issues](#)

[Children Diagnosed With Cancer: Returning to School](#)

[Children Diagnosed With Cancer: Late Effects of Cancer Treatment](#)

Health Professionals Associated With Cancer Care

Talking With Your Doctor (also in Spanish)

Coping with cancer

[After Diagnosis: A Guide for Patients and Families](#) (also in Spanish)

[Family and Medical Leave Act \(FMLA\)](#) (also in Spanish)

[Nutrition for Children With Cancer](#) (also in Spanish)

What Happened to You, Happened to Me (children's booklet)

When Your Brother or Sister Has Cancer (children's booklet)

When Your Child's Treatment Ends: A Guide for Families (booklet)

Cancer treatment information

[A Guide to Cancer Surgery](#) (also in Spanish)

[A Guide to Chemotherapy](#) (also in Spanish)

[Understanding Radiation Therapy: A Guide for Patients and Families](#) (also in Spanish)

[Clinical Trials: What You Need to Know](#) (also in Spanish)

Fertility and Women With Cancer

Fertility and Men With Cancer

[Second Cancers Caused by Cancer Treatment](#)

[Stem Cell Transplant \(Peripheral Blood, Bone Marrow, and Cord Blood Transplants\)](#) (also in Spanish)

Cancer treatment side effects

Caring for the Patient with Cancer at Home: A Guide for Patients and Families (also in Spanish)

Nausea and Vomiting

Guide to Controlling Cancer Pain (also in Spanish)

Anemia in People With Cancer

Fatigue in People With Cancer

Books

The following books are available from the American Cancer Society. Call us at 1-800-227-2345 or visit our bookstore online at cancer.org/bookstore to find out about costs or to place an order.

For family and friends of the child

American Cancer Society Complete Guide to Family Caregiving, Second Edition

Because... Someone I Love Has Cancer (kids' activity book)

Jacob Has Cancer: His Friends Want to Help (coloring book for a child with a friend who has cancer)

For the child with cancer

Imagine What's Possible: Use the Power of Your Mind to Take Control of Your Life During Cancer (grades 4 through 6)

Let My Colors Out (picture book for ages 5 to 10)

The Long and the Short of It: A Tale About Hair (ages 7 and up)

National organizations and websites*

Along with the American Cancer Society, other sources of patient information and support include:

American Childhood Cancer Organization (formerly Candlelighters)

Toll-free number: 1-855-858-2226

Website: www.acco.org

Offers information for children and teens with cancer, their siblings, and adults dealing with children with cancer. Also offers books and a special kit for children newly diagnosed with cancer, as well as some local support groups.

Children's Oncology Group (COG)

Website: www.childrensoncologygroup.org

Provides key information from the world's largest organization devoted to childhood cancer research to help support children and their families from the time of diagnosis, through treatment, and beyond. Also has a searchable database to find the COG center closest to you.

CureSearch for Children's Cancer

Toll-free number: 1-800-458-6223

Website: www.curesearch.org

Provides up-to-date information about childhood cancer from pediatric cancer experts. Has sections on the website for patients, families, and friends to help guide them on how to support the child with cancer.

National Cancer Institute

Toll-free number: 1-800-4-CANCER (1-800-422-6237)

TTY: 1-800-332-8615

Website: www.cancer.gov

Provides accurate, up-to-date information about cancer for patients and their families, including clinical trials information. Offers a special booklet for teen siblings of a child with cancer at: www.cancer.gov/cancertopics/when-your-sibling-has-cancer.

National Children's Cancer Society, Inc.

Toll-free number: 1-800-5-FAMILY (1-800-532-6459)

Website: www.children-cancer.org

Services include an online support network for parents of children with cancer, educational materials, and financial assistance for treatment-related expenses.

National Dissemination Center for Children with Disabilities (NICHCY)

Toll-free number: 1-800-695-0285 (also for TTY)

Website: www.nichcy.org

Provides information about disabilities and disability-related issues for families, educators, and other professionals.

Websites for children and teens

Starlight Children's Foundation

Toll-free number: 1-800-315-2580

Website: www.starlight.org

Website has animated stories and interactive programs to teach kids about chemo and procedures that may be done in the hospital; also has videos specifically for teens and provides a safe, monitored online support group for teens with cancer.

Group Loop (a subsite of the **Cancer Support Community** just for teens)

Toll-free number: 1-888-793-9355

Website: www.grouploop.org

An online place for teens with cancer or teens who know someone with cancer to connect with other teens – away from the pressures of classes, responsibilities, and treatment schedules. Has online support groups, chat rooms, information, and more.

SuperSibs! powered by Alex's Lemonade Stand

Toll-free number: 1-866-333-1213

Website: www.supersibs.org

Supports, honors, and recognizes 4- to 18-year-old brothers and sisters of children diagnosed with cancer so they may face the future with strength, courage, and hope. Alex's Lemonade Stand is restarting SuperSibs in 2014 so there may be some delays with resuming support services.

**Inclusion on this list does not imply endorsement by the American Cancer Society.*

No matter who you are, we can help. Contact us anytime, day or night, for information and support. Call us at **1-800-227-2345** or visit www.cancer.org.

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