Wilms' Tumor

Wilms' tumor is a rare kidney cancer that primarily affects children. Also known as nephroblastoma, Wilms' tumor is the most common cancer of the kidneys in children. Wilms' tumor most often affects children ages 3 to 4 and becomes much less common after age 5.

Wilms' tumor most often occurs in just one kidney, though it can sometimes be found in both kidneys at the same time.

Improvements in the diagnosis and treatment of Wilms' tumor have improved the prognosis for children with this disease. The outlook for most children with Wilms' tumor is very good.

Wilms' tumor doesn't always cause signs and symptoms. Children with Wilms' tumor may appear healthy, or they may experience:

- Abdominal swelling
- An abdominal mass you can feel
- Abdominal pain
- Fever
- Blood in the urine

When to see a doctor

Make an appointment with your child's doctor if you notice any signs or symptoms that worry you. The signs and symptoms associated with Wilms' tumor aren't specific to the condition and are much more likely to be caused by something else.

t's not clear what causes Wilms' tumor.

Doctors know that cancer begins when cells develop errors in their DNA. The errors allow the cells to grow and divide uncontrollably and to go on living when other cells would die. The accumulating cells form a tumor. In Wilms' tumor, this process occurs in the kidney cells.

In a small number of cases, the errors in DNA that lead to Wilms' tumor are passed from parents to children. In most cases, there is no known connection between parents and children that may lead to cancer.

Factors that may increase the risk of Wilms' tumor include:

- **Being black.** Black children have a slightly higher risk of developing Wilms' tumor than do children of other races. Children of Asian descent appear to have a lower risk than do children of other races.
- **Having a family history of Wilms' tumor.** If someone in your child's family has had Wilms' tumor, then your child has an increased risk of developing the disease.

Wilms' tumor occurs more frequently in children with certain abnormalities present at birth, including:

- **Aniridia.** In this condition the iris — the colored portion of the eye — forms only partially or not at all.
- **Hemihypertrophy.** A condition that occurs when one side of the body is noticeably larger than the other side.
- **Undescended testicles.** One or both testicles fail to descend into the scrotum (cryptorchidism).
- **Hypospadias.** The urinary (urethral) opening is not at the tip of the penis, but is on the underside.

Wilms' tumor can occur as part of rare syndromes, including:

- **WAGR syndrome.** This syndrome includes Wilms' tumor, aniridia, abnormalities of the genitals and urinary system, and mental retardation.
- **Denys-Drash syndrome.** This syndrome includes Wilms' tumor, kidney disease and male pseudohermaphroditism, in which a boy is born with testicles but may exhibit female characteristics.
- **Beckwith-Wiedemann syndrome.** Signs of this syndrome include abdominal organs that protrude into the base of the umbilical cord, a large tongue (macroglossia) and enlarged internal organs.

To diagnose Wilms' tumor, your child's doctor may recommend:

- **A physical examination.**The doctor will look for possible signs of Wilms' tumor.
- **Blood and urine tests.**Blood tests can't detect Wilms' tumor, but they can provide your child's doctor with an overall assessment of your child's health.
- **Imaging tests.** Imaging tests that create images of your child's kidneys can help your doctor determine whether your child has a kidney tumor. Imaging tests may include ultrasound, computerized tomography (CT) and magnetic resonance imaging (MRI).
- **Surgery.** If your child has a kidney tumor, your doctor may recommend removing the tumor or the entire kidney to determine if the tumor is cancerous. The removed tissue is analyzed in a laboratory to determine whether cancer is present and what types of cells are involved. This surgery may also serve as treatment for Wilms' tumor.

Staging

Once your child's doctor has diagnosed Wilms' tumor, he or she works to determine the extent (stage) of the cancer. Your child's doctor may recommend a chest X-ray, chest CT scan and bone scan to determine whether the cancer has spread beyond the kidneys.

The doctor assigns your child's cancer a stage, which helps determine the treatment options. The stages of Wilms' tumor are:

- **Stage I.** The cancer is found only in one kidney and generally can be completely removed with surgery.
- **Stage II.** The cancer has spread to the tissues and structures near the affected kidney, such as fat or blood vessels, but it can still be completely removed by surgery.
- **Stage III.** The cancer has spread beyond the kidney area to nearby lymph nodes or other structures within the abdomen, and it may not be completely removed by surgery.
- **Stage IV.** The cancer has spread to distant structures, such as the lungs, liver, bones or brain.
- **Stage V.** Cancer cells are found in both kidneys.

Treatment for Wilms' tumor usually involves surgery and chemotherapy. But treatments may vary by the stage of the cancer. Because this type of cancer is rare, your doctor may recommend that you seek treatment at a children's cancer center that has experience treating this type of cancer.

Surgery to remove all or part of a kidney

Treatment for Wilms' tumor usually begins with surgery to remove all or part of a kidney (nephrectomy). Surgery is also used to confirm the diagnosis, since the tissue removed during surgery is sent to a laboratory to determine whether it is cancerous.

Surgery for Wilms' tumor may include:

- **Removing part of the affected kidney.** Partial nephrectomy involves removal of the tumor and part of the kidney tissue surrounding it. Partial nephrectomy may be an option if your child's cancer is very small or if your child has only one functioning kidney.
- **Removing the affected kidney and surrounding tissue.** In a radical nephrectomy, doctors remove the kidney and surrounding tissues, including the ureter and adrenal gland. Neighboring lymph nodes also may be removed. The remaining kidney can increase its capacity and take over the entire job of filtering the blood.
- **Removing all or part of both kidneys.** If your child's cancer affects both kidneys, the surgeon works to remove as much cancer as possible from both kidneys. In a small

number of cases, this may mean removing both kidneys, which means your child would then undergo kidney dialysis. A kidney transplant may be an option so that your child will no longer need dialysis.

Chemotherapy

Chemotherapy uses powerful drugs to kill cancer cells throughout the body. Chemotherapy treatment usually involves a combination of drugs that work together to kill cancer cells. Chemotherapy can be administered through a vein or taken in pill form.

What side effects your child experiences will depend on which drugs are used. Common side effects include nausea, vomiting, loss of appetite, hair loss and frequent infections. Ask your child's doctor what side effects may occur during treatment, and if there are any potential long-term complications.

Chemotherapy may be used before surgery to shrink tumors and make them easier to remove. Chemotherapy may be used after surgery to kill any cancer cells that may remain in the body. Chemotherapy may also be an option for children whose cancers are too advanced to be removed completely with surgery.

For children who have cancer in both kidneys, chemotherapy is administered before surgery. This may make it more likely that surgeons can save at least one kidney in order to preserve kidney function.

Radiation therapy

Radiation therapy uses high-energy beams, such as X-rays, to kill cancer cells.

During radiation therapy, your child is positioned on a table and a large machine moves around your child, precisely aiming energy beams at the cancer. Possible side effects include nausea, diarrhea, fatigue and sunburn-like skin irritation.

Radiation therapy may be used after surgery to kill any cancer cells that weren't removed during the operation. Radiation therapy may also be an option to control cancer that has spread to other areas of the body, depending on where the cancer has spread.

Clinical trials

Your child's doctor may recommend participating in a clinical trial. These research studies allow your child a chance at the latest cancer treatments, but they can't guarantee a cure.

Discuss the benefits and risks of clinical trials with your child's doctor. The majority of children with cancer enroll in a clinical trial. However, enrollment in a clinical trial is up to you and your child.