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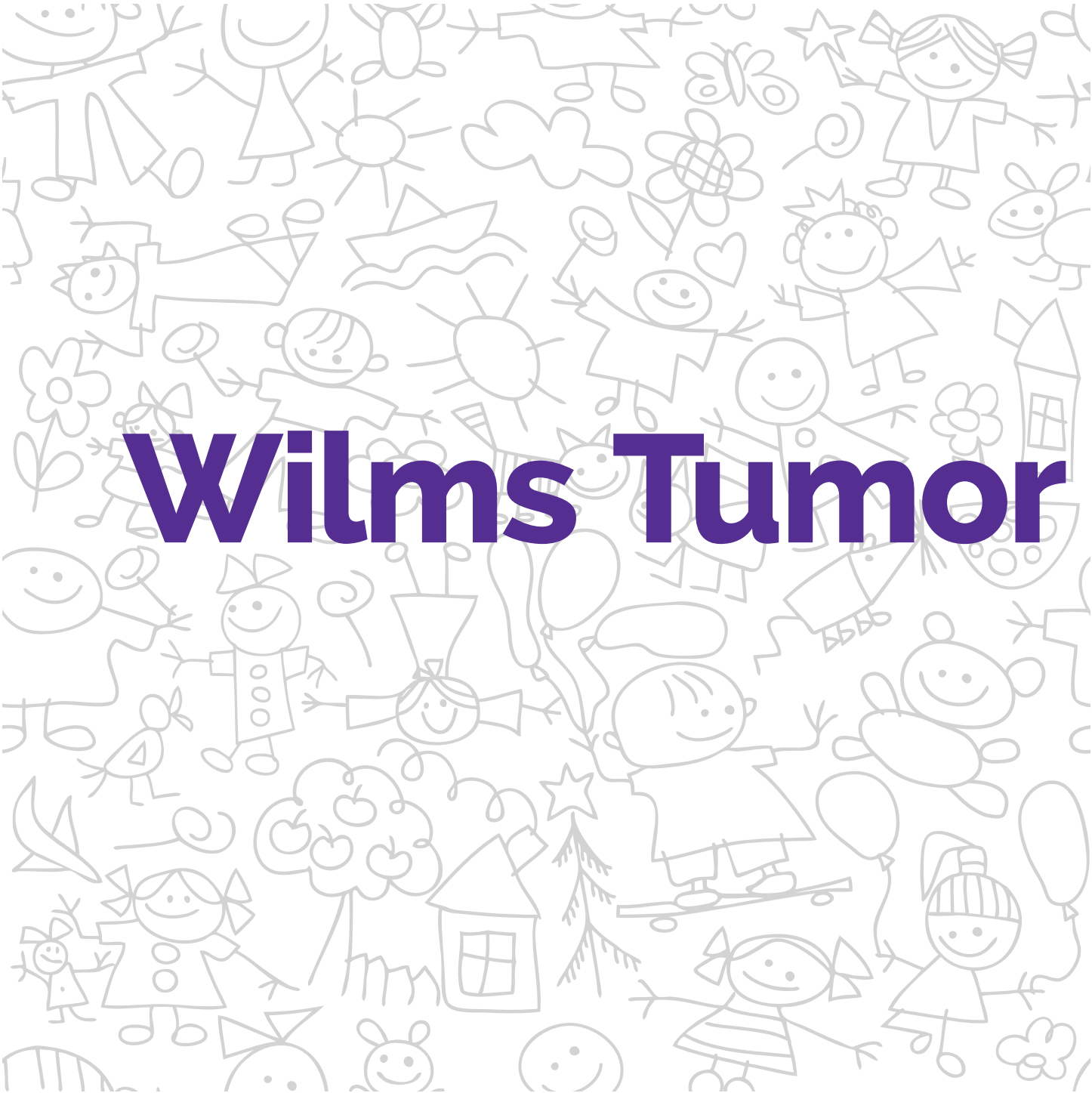
A Handbook for Families



Wilms Tumor

ONCOLOGY SERIES

WILMS TUMOR
A HANDBOOK FOR FAMILIES

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ABOUT THIS COVER

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WHAT IS WILMS TUMOR?

Wilms tumor is a cancer that develops from kidney cells. It is the most common childhood cancer of the kidneys. Wilms tumor can arise anywhere within the kidney. Rarely, Wilms tumor occurs outside of the kidney.

Not all of the kidney tumors found in children are Wilms tumors. Rhabdoid tumors and clear-cell sarcomas of the kidney are occasionally found in children. They are both more difficult to treat than Wilms tumor.

WHAT ARE SOME OF THE SIGNS AND SYMPTOMS OF WILMS TUMOR?

The most common sign of Wilms tumor is an enlarged or bloated belly in an otherwise healthy-looking child. Usually a caregiver notices the child's enlarged belly or feels a mass in the belly while bathing or dressing the child. Some other general signs and symptoms may be belly pain, tiredness, blood in the urine (usually found only by a urine test), and occasionally fever. One-fourth of the children diagnosed with Wilms tumor also have high blood pressure.

WHAT CAUSES WILMS TUMOR?

There is no answer at this time as to the cause of Wilms tumor. We do know, however, that it is not contagious or "catching." No behaviors or lifestyle habits, including those practiced during pregnancy, have been associated with the development of Wilms tumors. Cancer is not caused by anything the child ate or didn't eat (such as vegetables or vitamins).

WHO GETS WILMS TUMOR?

Wilms tumors account for 7% of all childhood cancers. Approximately 500 cases of Wilms' tumor are diagnosed each year in the United States. It is slightly more common among African-American children than among Caucasian children, and it is slightly more common among females than males. Most of the children (78%) diagnosed with Wilms tumor are between 1 and 5 years of age. The most common age at diagnosis is 3–4 years. It is rare in children over 10 years of age and less than 6 months of age.

IS WILMS TUMOR INHERITED?

Cancer may be caused by abnormalities in the genes, but only some forms are inherited. Studies have shown that 15%–20% of all Wilms tumors are inherited (one or more family members with a history of the disease). It appears that children who have the disease in both kidneys and who are younger at the time of diagnosis are more likely to have a genetic predisposition to develop Wilms tumor. The presence of Wilms tumor in other family members or the presence of intellectual disabilities or other birth defects in

the child is another indication that the child may be genetically predisposed to developing Wilms tumor. If your child's doctor suspects that your child has an inherited type of tumor, examination of family members and genetic counseling are available.

WHAT IS METASTASIS?

Metastasis refers to the spread of cancer from its original location (primary site) to other parts of the body. Wilms tumors may metastasize, or spread, through the blood system or the lymphatic system (tissues and cells that help to fight infection). The most common site of metastasis is the lungs. Wilms tumors sometimes spread to the liver, other kidney, brain, or bones.

WHAT IS STAGING?

Staging is the process of determining the extent of the disease at the time of diagnosis. The method of treatment proposed for your child will depend upon the stage of the disease at the time of diagnosis. Staging for Wilms tumor is done by the surgeon when the tumor is removed and by X rays and scans to look for metastatic disease.

The following staging system is recommended by the National Wilms Tumor Study Group. It is the major staging system currently used for Wilms tumor.

Stage I The tumor is only in the kidney, and the capsule around the kidney is not broken. There is no metastasis. The tumor can be removed by surgery.

Stage II The tumor extends outside the kidney and is pushing against the capsule that surrounds the kidney. The tumor has spread into the blood vessels outside the kidney. No spread can be detected at or beyond the edges of the tumor after surgery. The capsule that surrounds the kidney may have broken as it was being surgically removed, spilling cancer cells into the area outside the kidney.

Stage III The tumor has extended beyond the capsule of the kidney but is only in the abdomen. Any of the following conditions may exist:

- The tumor is found in the lymph nodes near the kidney.
- The tumor has spilled into the peritoneum (the membrane lining the walls of the abdomen or belly cavity and enclosing organs inside the abdomen) before or during surgery, or by growth into the peritoneal surface.
- Parts of the tumor are on the peritoneum.
- Tumor cells are on the edges of the tumor after its removal and in the tissue surrounding the area where the tumor had been in the body.
- The surgeon is not able to remove the entire tumor because it has spread into the surrounding area and has grown into important body structures that cannot be removed (biopsy only).

Stage IV The tumor has spread beyond the areas described in Stage III (for example, into the lungs,

liver, bone, brain, or distant lymph nodes).

Stage V Both kidneys have a tumor. Each kidney is staged according to the above criteria, based on the extent of disease before and after surgery.

WHAT TESTS AND PROCEDURES WILL MY CHILD NEED?

To diagnose Wilms tumor and determine the extent of your child's disease, a number of tests and procedures are necessary, including many of the following.

ULTRASOUND

This test uses high-frequency sound waves to look at internal body organs or tumors. It can help detect tumors in the lymph nodes or abdomen. It is painless and involves no radiation. If the abdomen is being scanned, the child is not allowed to eat or drink anything for 4 hours before the test but can resume a normal diet after the scan.

COMPUTERIZED AXIAL TOMOGRAPHY (CAT) SCAN

The CT or CAT scan is a computer-assisted X ray that shows very precise pictures of internal organs and tumors. The child must be able to lie absolutely still during the scan; some children require sedation. It may be necessary for the child to drink a liquid containing a flavorless dye that makes the pictures clearer. Sometimes a small amount of dye is injected into a vein. Generally, there are no side effects from either type of dye, although allergic reactions are possible.

X RAY

A chest X ray or chest CAT scan may be necessary to determine whether the tumor has spread to the lungs. An X ray is one form of radiation that can be used at low levels to produce an image of the body on film. Efforts will be made to minimize your child's exposure to X rays.

BLOOD TESTS

Blood tests are done to monitor the child's blood cells, body salts, and chemistries. A complete blood cell count (CBC) is useful in detecting a drop in the number of red blood cells (oxygen carriers), white blood cells (infection fighters), or platelets (cells that help blood clot properly). Blood chemistries such as BUN and creatinine monitor changes in kidney function. Liver function can be tested by looking at SGPT (ALT), SGOT (AST), and bilirubin. These tests are usually done at the time of diagnosis and are also checked throughout therapy to monitor the child's response to treatment.

CENTRAL VENOUS CATHETER (CVC)

A CVC is a permanent intravenous (IV) catheter used in patients receiving treatment for a chronic illness. CVC can be used for blood tests, medications, chemotherapy, blood products, and nutritional support, when needed. There are several types of CVCs, including tunneled CVCs (i.e., Broviac or Hickman), ports, and peripherally inserted central catheters (PICCs). Depending on the type of CVC, it may be inserted in the operating room while your child is under anethesia or the CVC might be placed using local (at the site only) pain medication. You and your child's medical team will jointly decide whether your child needs a CVC and which type.

HOW CAN WILMS TUMOR BE TREATED?

Three types of therapy are commonly used to treat Wilms tumor: surgery, radiation, and chemotherapy. The type of therapy chosen depends upon the extent of the disease. Your child's medical team will talk with you about the best treatment for your child.

SURGERY

Surgery is the first-line treatment for all stages of Wilms tumor. Surgery to remove the bulk of the tumor and the affected kidney is done at the time of diagnosis. In children whose tumor is too large, presurgery chemotherapy is given to shrink the tumor before removal. For children with Stage V Wilms tumor (both kidneys), the kidney with the largest tumor is removed. For some children, surgery is all that is needed to treat Wilms tumor.

CHEMOTHERAPY

Chemotherapy is medicine intended to kill tumor cells in the bloodstream or any tumor cells remaining elsewhere. Several chemotherapy medications are known to be effective in killing Wilms tumor cells, but no single chemotherapy medication can control this disease by itself. Most are given through a vein or CVC.

Your child's medical team will explain in detail the possible side effects of the specific chemotherapy medicines recommended for your child.

RADIATION

Radiation therapy is a special kind of X-ray treatment. Radiation treatment is very precise and is given in specially measured amounts by radiation-therapy experts. Radiation is often used together with chemotherapy.

The purpose of radiation therapy is to kill cells that may have escaped from the tumor. Wilms tumor cells are often very sensitive and easily killed by radiation. Usually children with Stage I or Stage II disease do not receive radiation. If radiation is necessary for your child, the radiation-therapy team will discuss with you exactly how the radiation will be given and how long the treatments will last.

In general, children experience very few side effects while they are getting radiation therapy. Some children may be more tired than usual or have decreased appetites. There can also be long-term side effects, which will be explained to you in detail.

HOW LONG WILL MY CHILD'S THERAPY LAST?

The length of treatment will depend on tumor staging. Chemotherapy treatment generally lasts 5–7 months. It involves weekly visits to the clinic for chemotherapy or blood tests, or both.

WHAT NEW METHODS OF TREATMENT ARE THERE?

The most recent published data reveal that more than 90% of children survive Wilms tumor. Because most children with Wilms tumor are cured of their disease, researchers are now looking at ways to fine-tune the treatment. Children with a good prognosis (favorable outcome) may require less treatment in the future. Children with unfavorable histology or with metastasis may require more intensive treatment. Effective treatments are being developed for the 10%–15% of children who have a recurrence of Wilms tumor. New studies will help us see if current genetic markers are associated with the aggressiveness of the tumor or how the tumor responds to treatment.

Most of the advances in the treatment of childhood cancer have been made through a process known as clinical trials. In clinical trials, the best-known (standard) treatment for a particular cancer is compared to a new (research) treatment that is believed to be at least as good as, and possibly better than, the standard treatment. Clinical trials allow the medical team to determine whether promising new treatments are safe and effective.

Participation in clinical trials is voluntary. Since clinical trials involve research into new treatment plans, all risks cannot be known ahead of time, and unknown side effects may occur. On the other hand, children who participate in clinical trials may be among the first to benefit from new treatment approaches. Before making a decision about your child's participation in a clinical trial, the risks and potential benefits should be discussed with your child's medical team.

More information about clinical trials is available at www.cancer.gov/cancertopics/factsheet/Information/clinical-trials. You can also call 800.4CANCER (800.422.6237).

WHAT ARE THE POTENTIAL LONG-TERM EFFECTS OF THERAPY?

The major concern in children with Wilms tumor is long-term function of the remaining kidney. The most current data show that only 0.25% of the children with the disease in only one kidney developed kidney failure. Children with the disease in both kidneys, and those who had a syndrome associated with Wilms tumor, had a higher risk of developing kidney failure.

Children who received radiation therapy for their disease are at risk for mild scoliosis, muscular hypoplasia (underdevelopment of the muscle), and fertility and pregnancy problems (increased numbers of spontaneous abortions, low birth-weight babies, premature babies, babies with congenital problems, and neonatal deaths).

A small number of children treated for Wilms tumor may develop congestive heart failure due to one of the chemotherapy drugs. Also, a very small number of children have developed a second cancer due to the chemotherapy given. Your child's treatment team can discuss these risks with you in more detail.

HOW CAN I WORK WITH MY CHILD'S HEALTHCARE TEAM?

Your child's care requires a team approach. As caregiver, you are a major part of the team—your input is important. Because you know your child better than anyone else, the medical staff will need your help to deliver comprehensive management for the disease.

It is important to communicate openly with your child's healthcare team. Be sure to question your child's medical team whenever there is anything you are not sure about. It helps to write down your questions when you think of them.

Here are some examples of questions to ask:

- **What** kind of cancer does my child have?
- **Has** the cancer spread beyond the primary site?
- **What** is the stage of the cancer, and what does that mean?
- **What** treatment choices are available?
- **What** treatment do you recommend, and why?
- **What** risks or side effects does the recommended treatment have?
- **What** should we do to prepare for treatment?
- **What** is my child's outlook for survival?
- **What** are the chances of a recurrence?

Use this space to write down some additional questions:

ARE MY FEELINGS NORMAL? WHAT CAN I DO ABOUT THEM?

Hearing that your child has cancer can be shocking and overwhelming. At first you may not believe it, or you may hope that the diagnosis is wrong. However, the changes you see in your child and the experience of being in the hospital and beginning treatment will no doubt confirm the reality of your child's situation.

Many family members feel that they are somehow responsible for their child's disease, or they feel guilty that they were not able to detect it sooner. The disease was not caused or triggered by anything anyone did to or fed to the child or by anything that happened during the pregnancy with the child.

In addition to shock and guilt, you and your family will probably feel anger and sadness. Even the youngest family members are likely to be affected. These feelings are normal, and each family member will express them in different ways and at different times. It can be very difficult to feel so many strong emotions at once. Talking honestly with one another about feelings, reactions, and questions will help everyone in the family.

It may seem difficult to talk to friends, family, or even medical staff, but venting your feelings can help you cope. Your child will benefit if family members continue to show their care through support and communication.

HOW CAN I HELP MY CHILD?

As a caregiver, you will often notice changes in your child during the treatment. These changes or symptoms are usually due to the treatment. They can make you feel even more helpless. It is essential always to remember that, in spite of changes on the outside, your child is still the same person on the inside. Hair loss and other changes in body appearance are temporary. They often bother the adults involved much more than the child or the child's siblings and friends. All of your feelings about what your child is going through during treatment must be balanced by remembering that treatment provides an opportunity to cure the disease and to have your child go on to a full and meaningful life.

It is important to reinforce to your child that nothing he or she did or said caused this disease. Telling your child that your anger or sad feelings are directed at the cancer and not at him or her will help keep your special relationship honest and close. Like you, your child will need someone with whom he or she can share feelings. Don't hesitate to ask your child to express his or her feelings, and don't be afraid to explain what is happening and why.

In spite of the disease, your child is still growing and learning. All children, sick and well, need love, attention, discipline, limits, and the opportunity to try new skills and activities. As you begin to learn the new, special needs of your child, it is important to remember that he or she still has all the needs and rights of any growing, developing person. Do not avoid using direct terms and explanations with your child. Children tolerate treatment better if they understand it and are allowed to be active decision makers whenever possible. The same is true for caregivers!

IS MY CHILD'S DIET IMPORTANT DURING THE TREATMENT?

Yes. We know from research that well-nourished children tolerate therapy better and have fewer treatment delays due to illness. It may be difficult for your child to resume normal eating habits while on therapy, so you will need to be flexible and creative. Often, numerous small meals are tolerated better than three large ones. Children are usually more interested in eating foods that they help prepare. It is important to include the child in the social activity of family meals even if full meals aren't eaten. Remember, nobody wins food fights—it is best not to force your child to eat.

Make sure that foods high in protein and carbohydrates are readily available. Multivitamins, medicines, and herbs should be approved by your doctor before you give them to your child because there may be an interaction between them and chemotherapy. A dietitian trained in the calorie and energy needs of children with cancer can offer you guidance. The medical staff can intervene if there is a nutritional problem.

CAN MY CHILD ATTEND SCHOOL DURING TREATMENT?

Your child's ability to attend school will depend on the intensity of the therapy and on the response to treatment. Some children tolerate chemotherapy and radiation better than others. Your child may not be able to attend school for extended periods due to treatment or hospitalization. However, it is important that your child keep up with his or her schoolwork. Talk to your child's regular school about arranging services, including help from a home tutor, until he or she is able to return to school. You also should discuss school attendance with your child's medical team. Many pediatric hospitals have school programs that enable children to attend school while hospitalized.

School is important because it helps children maintain social contact with their peers. Maintaining time with friends will be an important part of your child's recovery and will ease the adjustment when he or she returns to school. It is important for your child to return to school as soon as he or she is medically able to do so.

SUGGESTED READING

National Cancer Institute (2013). Wilms Tumor and Other Childhood Kidney Tumors. Retrieved from: http://www.cancer.gov/cancertopics/pdq/treatment/wilms/Patient

CureSearch for Children's Cancer(2011). Kidney Cancer in Children (Wilms Tumor): Just Diagnosed Information. Retrieved from: http://www.curesearch.org/Wilms-Tumor-in-Children-Just-Diagnosed-Information

IMPORTANT PHONE NUMBERS

NOTES

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