INFORMATION ABOUT WILMS’ TUMOR

WHAT IS WILMS’ TUMOR?
Wilms’ tumor is the most common type of kidney cancer in children. It occurs from infancy to age 15 but is most frequently seen in the first 5 years of life. The kidneys are two bean shaped organs which are responsible for filtering and removing waste products from the blood in the form of urine. A tube called the ureter connects the kidney to the bladder where urine stored until it passes out of the body.

WHAT CAUSES WILMS’ TUMOR?
The exact cause of Wilms’ Tumor is not known although certain risk factors have been identified as being associated with its occurrence. A risk factor is anything that increases a person’s chance of getting a disease. For Wilms’ Tumor, this includes certain birth defects or combinations of birth defects (genetic syndromes). Some of these birth defects include absence of the iris (colored portion of the eye), ambiguous genitalia, abnormal enlargement of one half of the body (also called hemihypertrophy), undescended testes, and rare syndromes such as Beckwith-Wiedeman syndrome, Denys-Drash syndrome, etc. Not all children with a risk factor will develop Wilms’ tumor and the majority of children who develop Wilms’ tumor have no identified risk factor. For those children affected with these genetic syndromes, it is recommended that they undergo abdominal ultrasound regularly to screen for Wilms’ Tumor.

WHAT ARE THE SYMPTOMS OF WILMS’ TUMOR?
The child often presents with an enlarged abdomen, which is usually noted by the caretaker while bathing or dressing the child. Other symptoms include abdominal pain, blood in the urine, increased blood pressure (hypertension) and fever. If a child experiences any of these problems, it is best for him or her to be seen by a physician.

HOW IS THE DIAGNOSIS OF WILMS’ TUMOR MADE?
A careful health history and physical examination will be performed as well as several tests done to prove the diagnosis of Wilms’ Tumor, determine the stage and to determine whether its spread to the lung, lymph nodes or liver.

A family history of cancer is generally asked, as well as the patient’s past illnesses and treatments.

The physical examination may reveal the presence of abdominal enlargement or a palpable mass in the flank area.

Special imaging test are requested to better locate and characterize the size of the tumor such as an abdominal ultrasound, a CT (Computerized Tomography) scan or an MRI (Magnetic Resonance Imaging).

Other tests that may be requested include a complete blood count, baseline liver and kidney function tests, serum calcium, screening for coagulation factors, a urinalysis, heart function tests and in suspected spread of disease to other parts of the body, a CT scan of the chest, MRI of the brain and a bone scan.

A piece of the tumor will be taken (a biopsy) and will be sent to a pathologist for microscopic examination and confirmation of the diagnosis of Wilms’ Tumor.

HOW DOES ONE STAGE WILMS’ TUMOR?
Staging refers to the process used to follow to find out if the cancer stays within the affected kidney or has spread to other parts of the body. Knowing the disease stage is important for treatment planning as well as knowing the child’s chance of cure.

Stage I: refers to the disease being confined to the kidney, which has been surgically removed and intact. There is no involvement of nearby tissues or blood vessels of the kidney

Stage II: the kidney was entirely removed surgically but there is involvement of nearby tissues and blood vessels of the kidney
Stage III: the disease kidney was not removed surgically. Other tissues are also involved such as the lymph nodes in the abdomen and pelvis, the peritoneum or tissue lining the abdominal cavity and other abdominal organs. This also includes cases wherein only a biopsy was done, or if there was spillage or potential spillage of tissue during surgery.

Stage IV: includes involvement of other organs such as the lungs, bone, brain, liver or lymph nodes outside of the abdomen and pelvis.

Stage V: refers to both kidneys being involved at diagnosis. Each kidney is also staged individually.

**How is the treatment planned and who are involved in the treatment?**

In planning the treatment for a child with Wilms' tumor, certain factors that impact on the chance of recovery are taken into consideration. This includes histology or the microscopic appearance of the tumor cells, tumor size, age of the child, whether or not the tumor was completely removed surgically, and whether there are any abnormal chromosomes or genes that may affect the chance of cure.

The pediatric oncologist will be working very closely with other experts such as a pediatric surgeon or urologist, a radiation oncologist as well as many other health care professionals such as oncology nurses, pharmacists, psychologists, social worker, etc. This is called multidisciplinary care and aims to address the many needs of the child as completely as possible.

**What are the available treatment options for Wilms' Tumor?**

The treatment options for children with Wilms' Tumor include the following:

Surgery is performed to remove the tumor as much as possible. The kidney involved with the tumor is completely removed as well as neighboring tissues and lymph nodes. This is called radical nephrectomy. In cases wherein both kidneys are involved, the less affected kidney undergoes a partial nephrectomy so that enough kidney tissue is left to maintain normal function. During surgery, the surgeon is likewise able to determine whether adjacent tissues, the blood vessels of the kidney or lymph nodes in the abdomen and pelvis or peritoneum are involved.

Chemotherapy is the use of drugs to kill cancer cells. It may be given by mouth or injected through a vein or muscle (systemic therapy). For children with Wilms' tumor, a combination of different drugs known to kill Wilms’ tumor cells is given. The drugs may be given before (neoadjuvant) and/or after (adjuvant) surgery.

Radiation therapy uses high-energy X-rays or other radiation energy to kill cancer cells. Radiation therapy is administered by a radiation oncologist.

Aside from these standard therapies, studies are currently being done using biologic therapy, wherein the patient’s immune system is used to fight cancer as is the use of very high doses of chemotherapy followed by the transplantation of blood forming stem cells (hematopoietic stem cell transplant).

The exact choice of treatment combinations (surgery, specific chemotherapeutic drugs, extent of radiation therapy and length of therapy depends on the histology (microscopic appearance of the tumor) and stage and will be explained to you by your pediatric oncologist.

This information is made possible through the efforts of the Philippine Society of Pediatric Oncology, Inc. (PSPO), a subspecialty society of the Philippine Pediatric Society (PPS). For details regarding the treatment of individual patients, it is strongly recommended that they confer with their pediatric oncologist.