Wilms tumor (nephroblastoma)

Patient and family information, brought to you by the Education Committee of APSA

Overview - "What is it?"

Wilms tumor is the most common cancer of the kidney seen in infants and children. It is made up of tissue that comes from immature cells of the kidney. Wilms tumor is named after Dr. Wilms, a pathologist who first described the tissues from which this tumor arises.

About 500 cases are diagnosed in the United States per year, representing a little more than 10% of all childhood cancers. The average age of the patient at presentation is three years of age and most commonly seen between six months to eight years of age.

Signs and Symptoms - "What symptoms will my child have?"

The most common presentation is a firm, large, non-painful abdominal mass, commonly felt by a parent during a bath or a pediatrician during a well-baby check. The child may also have high blood pressure or blood in his or her urine. Other findings may include weight loss, loss of appetite or decreased energy level.

Diagnosis - "What tests are done to find out what my child has?"

Physical exam: If the doctor is concerned that a child's abdomen is distended, he or she may request several imaging studies.

X-ray of the belly: may distinguish whether the distention is due to intestinal problems.

Ultrasound: may be performed if a mass in the abdominal organs, such as the kidney is suspected. This test can definitively pinpoint the origin of the mass and may show whether the mass has extended beyond the kidney or spread into surrounding tissues such as the lymph nodes or blood vessels. In addition, it is important to determine whether there is anything suspicious for cancer in the other kidney.

An ultrasound is a more sensitive test to see whether there is tumor in the veins next to the kidney. Knowledge of the extent of the tumor (one kidney vs both kidneys, outside tumor spread into the lymph nodes, lungs or blood vessels) is important.

Computed tomography (**CT) scan:** the CT will determine whether removal of the tumor is safe for your child ("tumor resectability"); sometimes, if the tumor is really big and surrounds organs and structures that are critical, its immediate removal may not be safe for your child.

Wilms tumors can spread to the lungs as well, so a CT of the lungs is typically performed.

- The spread of the tumor affects the type and order of treatments. For example, a tumor that is in one kidney only may be treated by surgery alone, but if there is spread of the tumor to the other kidney or other organs, chemotherapy may be the first treatment that should be done to get the best outcomes for your child.

Labs and tests: Bloodwork will be drawn to determine the child's general health including the blood count (red cell count) and the function of the kidney. The spread of the tumor determines the "stage" of the tumor.

Conditions that mimic this condition: Other tumors or growths arising from the kidney and other abdominal organs such as the liver can mimic Wilms tumor. Therefore, X-rays are very important to confirm the diagnosis.

Treatment - "What will be done to make my child better?"

Surgery: The type of operation needed for Wilms tumor depends on the extent or spread of the tumor. If the tumor is all within the kidney and no spread is seen outside of the kidney, removal of the involved kidney may be the only treatment needed. If the tumor is in both kidneys or has spread into the vena cava (large vein next to the kidney) or the lungs, the initial treatment may chemotherapy and not surgery. In these cases, a biopsy of the tumor is needed to see what type of tumor (aggressive or not aggressive) exists. At the same time, a catheter is placed into the central vein of your child to allow infusion of medications that fight cancer (chemotherapy). In situations where the tumor is aggressive or it goes beyond the confines of one kidney, chemotherapy has been shown to make the overall outcome better for the child. Once the tumor shrinks, then definitive surgery to remove the main tumor is performed.

Preoperative preparation: As already mentioned, X-rays to determine whether the tumor is resectable or whether primary removal of the tumor is the right thing for your child. Depending on the complexity of the surgery, preparation for the possibility of blood transfusion may be done.

Postoperative care: Your child may require some days in the Intensive Care Unit, depending on the extent of the surgery.

Important miscellaneous issues: The prognosis of Wilms tumor depends on many factors including your child's age, the size (weight) of the tumor, the aggressiveness of the tumor cells as determined by their properties (will be tested by the pathologists) and tumor spread.

Depending on any and/or a combination of these factors, your child may need surgery alone; a combination of surgery, chemotherapy and radiation; or even stem cell transplant. Your child's surgeon and cancer doctor (oncologist) will discuss these options with you as they become aware of the information about the tumor and its properties.

Medicine: The medicines that will be part of your child's chemotherapy regimen will be discussed with you by his or her oncologists. Each medicine has its side effects that can affect your child in both the short term and long term. Be sure that you are aware of these issues during the discussion.

Risks/Benefits: Risks of surgery include bleeding (which may require transfusion), infection and damage to surrounding organs.

Home Care - "What do I need to do once my child goes home?"

Diet: Usually the child can be on a regular age-appropriate diet when he or she goes home. However, during chemotherapy, the child may lose interest in eating. To keep up his strength, sometimes the child may need supplementation of nutrition using a feeding tube (tube that goes from the nose to the stomach) or a venous catheter (PICC® or central line). This issue should also be addressed by the oncologist. The care at home will, to some extent, depend on the type of surgery that was needed and the age of the child when the surgery was done.

Activity: If your child had a biopsy only, regular activity can be resumed about two days after surgical procedure. If he or she had a large incision (could be up-and-down or side-to-side incision), then he/she may need to decrease vigorous activity (jumping, running, climbing, contact sports) for about 3-4 weeks while he or she is healing.

Wound care: Dressings will be removed in 48 hours. No further dressing is required. Usually, the wound has to be kept dry for about 2-3 days. After this, the child can shower, but he or she should not soak the wound for about one week after surgery.

Medicines: Pain medication will be given. Narcotics may be needed at first, but by the time of discharge are typically not needed. Tylenol alone is typically the only thing required. Constipation is a significant side effect from narcotics. Stool softeners should be given to prevent this. In general, ibuprofen (Motrin® or Advil®) should not be given to patients with only one kidney.

What to call your doctor: A call to the surgeon should be made if there is worry about infection (unexplained fevers, redness and drainage of the wound(s)), vomiting or inability to have bowel movements. Your child's oncologist will likewise give you instructions about when to call their clinic.

Follow-up: A wound check is often performed in 2-3 weeks after surgery. Often, the child's oncologist can also provide follow up of the wound as well.

Long Term Outcomes - “Are there future conditions to worry about?”

Most children with Wilms tumor can be cured and do very well. However, outcomes depend on the characteristics of individual tumor and spread (stage) of the tumor. Long-term outcomes may also be affected by side effects of the chemotherapy. It is best to have these individualized discussions with your child’s oncologist and surgeon.

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