Signs of Wilms Tumor

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Wilms tumor, also called "nephroblastoma," is most common in boys and girls between one and four years of age. Black children, especially girls, seem to be more susceptible than white children of either sex. The most common sign by far is the presence in an otherwise healthy child of a painless lump in the side, not the middle, of the belly—that is, the flank.

Classically, it is first noted by a grandmother or aunt who comes to visit and while bathing the child first feels a mass. Mothers very often do not notice the gradual increase in size, and assume that this is just another part of a big belly, so often seen in healthy, chubby children. Abdominal pain, fever or bloody urine are less common first signs, or high blood pressure may be found during a routine well-baby visit to the pediatrician.

Very uncommon as the sole presenting sign is a varicocele in boys; that is, swelling of the veins near the testicle. It is due to pressure on the great vein of the abdomen (the vena cava) causing back-up of normal drainage.

Sometimes Wilms tumor occurs in children who have had other malformations or abnormality syndromes. Here are a few of the most common ones:

OVERGROWTH ABNORMALITIES

These in turn can be subdivided into **hemihypertrophy** and the **Beckwith-Wiedemann Syndrome** (BWS). Hemihypertrophy means that one side of the body—in whole or in part—is larger than the other side. Thus, one whole leg or arm may be longer and bigger around than the other, but even smaller parts like one side of the face may be asymmetrical. The BWS is more complex. It is characterized by a large tongue that may protrude from the mouth, a hernial sac in the belly button (called an omphalocele), and low blood sugar levels. Children with the BWS may exhibit enlargement of other organs such as the liver and pancreas. Hemihypertrophy may also be seen. They can develop cancers other than Wilms tumor (for example malignant growths in the liver and the gland above the kidney which therefore is called the suprarenal gland).

THE WAGR SYNDROME

The W stands for **W**ilms tumor, the A for **A**niridia, the G for abnormalities of the **G**enito-urinary organs, and the R for **R**etardation. Aniridia is the absence of the iris of the eye present at birth. The genito-urinary malformations include major anatomic maldevelopments. These include mixed development of the sexual organs making identification of sex uncertain, undescended testicle, or severe hypospadius. In this latter condition the urinary stream issues not at the tip of the urethra but from an abnormal orifice along the shaft of the penis or even higher than that; for instance, in the perineum. The R in mental Retardation completes the acronym.

DENYS-DRASH SYNDROME

This rare anomaly affects boys more than girls. It varies in complexity and severity, but abnormally developed kidneys that do not function normally are the principal feature. The children may be mentally retarded and may share other characteristics of the WAGR syndrome including ambiguous genitalia. They are also at higher risk than other children for developing Wilms tumors. Those with pronounced kidney problems rarely survive adolescence.

SUMMARY

Children with any of the rare syndromes described require careful follow-up in specialty clinics. This is needed not only for the other problems that make up these complicated anomalies, but also in order to detect a tumor when it is still small. Otherwise, parents need to be alert to any sign of a growing lump anywhere in the child's body; for instance, by feeling for one in the abdomen while changing a diaper or while bathing the child.