New Guidelines for Treating Wilms Tumor, A Childhood Cancer

New guidelines for treating Wilms Tumor share information for earlier, safer diagnosis and treatment in an effort to reduce disparities.

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The National Comprehensive Cancer Network® (NCCN®) today published its first-ever treatment guidelines for children with a solid tumor cancer type in the United States. The NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®) for Wilms Tumor (Nephroblastoma) contain evidence-based expert consensus recommendations for how to maximize benefits and minimize side effects for managing a rare type of kidney cancer that typically occurs in children age eight years or younger. These guidelines contain crucial information, particularly for care providers who treat a full spectrum of pediatric tumors, hematologic malignancies, and other blood disorders (such as sickle cell), due to the relative infrequency of all of these conditions. The guidelines also include important diagnostic information for primary care pediatricians to help ensure this cancer is caught early enough to benefit from high curability rates, and managed in a way that reduces side effects.

“Wilms tumor is rare, but highly treatable,” says Daniel M. Green, MD, of St. Jude Children’s Research Hospital, Vice-Chair of the NCCN Guidelines® Panel for Wilms Tumor. “It’s important that we make sure these guidelines and knowledge of the signs and symptoms of Wilms tumor reach primary care physicians and pediatricians so they can help catch it early and save lives.”

“Keeping up with all of the current literature for the broad array of children’s oncologic and hematologic malignancies can be daunting. The NCCN Guidelines for Wilms Tumor provide compiled, annotated, and vetted recommendations for diagnosis, treatment, and follow up—in keeping with NCCN’s goals for streamlining and improving care so that patients can live better lives,” said Elizabeth Mullen, MD, of Dana-Farber/Boston Children’s Cancer and Blood Disorders Center, another Member of the NCCN Guidelines Panel for Wilms Tumor. “People say Wilms is a ‘good cancer’ because it has a 90 percent cure rate but that still means too many children don’t survive. I won’t feel okay until we get that cure rate to 100 percent—with optimization of long-term wellness, fertility, and psycho-social wellbeing.”

“The treatment of Wilms tumor is guided by the results of a series of sequential clinical trials that started back in the 1960s,” explained Frank M. Balis, MD, Professor of Pediatrics, Louis and Amelia Canuso Family Endowed Chair for Clinical Research in Oncology, Children’s Hospital of

Philadelphia/Abramson Cancer Center at the University of Pennsylvania, who chairs the NCCN Guidelines Panel for Wilms Tumor. “Treatment approaches have been refined through these clinical trials, making treatment selection more and more complex and nuanced. Management should be determined by clinical and biological risk factors, the presence of unilateral (single kidney) versus bilateral (both kidneys) disease, and patient response to treatment. The NCCN Guidelines for Wilms Tumor present clinicians with the latest evidence for the management of children with Wilms tumor based on their individual clinical and molecular profile.”

Wilms tumor accounts for five percent of all childhood cancers and typically occurs between ages one to five. Children often have abdominal swelling and/or a mass with or without other symptoms; however, they often appear healthy, which can delay diagnosis. Wilms tends to be more common among African-American children and is often caught in more advanced disease stages for this population.[1],[2] In rare cases, children with a number of genetic predisposition syndromes may get Wilms tumor; therefore, these children should be screened for Wilms tumor.

“One of the most common ways that Wilms tumor is diagnosed in a child is through detection of an otherwise asymptomatic abdominal mass during a routine well child care exam with their pediatrician,” explained Dr. Mullen. “Now that some parents and guardians have had to delay these appointments, or hold them virtually due to the COVID-19 pandemic, there is concern about the potential of an increase in higher-stage diagnoses.”

The guidelines also contain a warning for primary care physicians to be gentle if they feel a mass in the kidney area, especially if the children appear to be otherwise healthy. Wilms tumor(s) can rupture if touched with too much force, causing the cancer to spread within the abdomen or elsewhere.

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