Wilms' Tumor and Other Pediatric Renal Masses

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• Wilms' tumor • Children • MR imaging

Wilms' tumor, or nephroblastoma, is the most common primary malignant renal tumor in children.[1–3] It tends to occur in children younger than 5 years of age, with prognosis varying with pathology, extent of disease, and tumor resectability. In this article, we review the role of MR imaging in the evaluation of Wilms' tumor and various other renal neoplasms that can occur in the pediatric population. MR and clinical features that are helpful in differentiating among these lesions are emphasized.

TECHNIQUE

Imaging Parameters

Lesion detectability depends on the signal-to-noise ratio, spatial resolution, and contrast resolution. These parameters vary with receiver coil size, slice thickness, field of view, matrix size, and number of acquisitions. The smallest coil that fits tightly around the body part being studied should be used to optimize signal-to-noise ratio and spatial resolution. A head coil usually is adequate in infants and small children, whereas a phased array or whole-body coil is usually needed for larger children and adolescents.[4,5] Surface coils can be useful in the evaluation of superficial structures, such as the spine, but the drop-off in signal strength with increasing distance from the center of the coil limits the value of these coils in the evaluation of deeper abdominal structures.

MR imaging of the primary tumor should be performed in at least two planes, usually coronal and axial. Slice thickness varies from 3 to 6 mm, depending on patient size. The field of view can have a square or rectangular shape. A square shape is used when the body part being examined fills the field of view. An asymmetric rectangular field of view is ideal for body parts that are narrow in one direction, such as the abdomen in a thin patient. To shorten imaging time, MR imaging examinations are obtained with 128 to 192 phase-encoding steps and one or two signal acquisitions.

Imaging Sequences

MR imaging of the primary tumor should be acquired with (1) T1-weighted, (2) T2-weighted, (3) gradient echo, and (4) gadolinium-enhanced, fat-saturated T1-weighted sequences.[4–7]

Pulse sequences

Spin-echo sequence T1-weighted sequences (short TR, short TE) provide excellent contrast between soft-tissue structures and fat and help in tissue characterization (ie, fluid, fat, or blood). Fat-suppressed T1-weighted images are useful to improve conspicuity of diseased tissues. T2-weighted sequences (long TR, long TE) provide excellent contrast between tumor and adjacent soft tissues and are useful for tissue characterization. The T2-weighted sequences can be acquired with either conventional or fast/turbo techniques. The latter are advantageous because they shorten imaging time. The fast spin echo techniques, however, may result in some loss of contrast between fat and other high signal intensity tissues or fluid. They should be used with fat-suppression techniques to increase lesion conspicuity. Two basic methods of fat suppression are widely

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available: short-tau inversion-recovery and radiofrequency presaturation of the lipid peak.

Most lesions have low signal intensity on T1-weighted images and high signal intensity on T2-weighted images. The T1 signal can increase if the lesion contains fat, blood, proteinaceous fluid, or cartilage. Gadolinium chelate enhancement also results in high T1 signal intensity. A low T2 signal intensity is seen with mineralization (calcification), hemosiderin and other blood products, iron oxide, and fibrosis.

Gradient-echo or time-of-flight images result in high signal in flowing blood and are useful for detecting flow in vascular structures.[7] By comparison, flowing blood appears as a flow void or decreased signal within the vessel lumen on spin echo sequences. GE imaging is most effective in cooperative children who can suspend respiration, but it can be used in children of any age. The evaluation of blood flow with the GE sequence requires the use of technical parameters that are tailored for vascular imaging. The short imaging time required to obtain individual images on GE sequences also provides the ability to dynamically monitor contrast enhancement after an injection of gadolinium compounds.

Chemical shift imaging is an optional method that is helpful to detect and characterize lesions suspected of containing fat. This technique uses GE images obtained in phase and out of phase to exploit differences in precessional frequencies of fat and water. The presence of fat and water within a voxel results in phase cancellation and decreased signal intensity on out-of-phase images. The shortest possible TE should be used for out-of-phase imaging to reduce T2* effects.

Contrast-enhanced imaging

Gadolinium chelates are extracellular contrast agents that cause T1 shortening of tissues.[4–6] The usual dose is 0.1 mmol/kg. T1-weighted fat-suppressed sequences after the administration of intravenous gadolinium chelates can help define areas of necrosis and cyst formation and may improve contrast between tumor and normal tissues.

Optimizing image quality

Voluntary motion can be minimized or eliminated by the use of sedation, whereas physiologic motion and its resultant artifacts—ghosting and blurring—can be suppressed by the use of fast imaging techniques, such as GE, fast spin echo and single-shot fast spin echo sequences, and signal averaging (ie, increasing the number of excitations).[4–7] Respiratory triggering can be used to reduce breathing artifact.

Conscious sedation Voluntary motion can be minimized or eliminated by the use of conscious sedation. Children older than age 6 usually cooperate for the MR examination after an explanation of the procedure and reassurance. It also helps to ensure that the patient is comfortable and free of pain and has an empty bladder. Conscious sedation is essential when imaging children younger than 6 years of age. Oral chloral hydrate is the most common sedative in children younger than 18 months. In children aged 18 months to 6 years, intravenous pentobarbital sodium is usually the drug of choice. Because it has a short half-life, propofol has been used in some centers; however, its use usually requires the assistance of an anesthesiologist.[8,9]

WILMS' TUMOR

Epidemiology

Wilms' tumor is the most common primary malignant renal tumor in children, accounting for 7% of all childhood cancers.[1–3,10,11] The annual incidence is 1 in 10,000 children younger than 15 years of age worldwide.[1–3] Approximately 450 to 500 new cases are diagnosed each year.[1,2,10] Wilms' tumor is more common in African Americans than in whites and is rare in Asians.[3,4,10,11] The tumor is equally common in girls and boys.[11] The mean age at diagnosis is 3.5 years.[2,3]

Risk Factors

Several congenital abnormalities (hemihypertrophy and sporadic aniridia) and syndromes, including Beckwith-Wiedemann (hemihypertrophy, omphalocele, macroglossia and visceromegaly), WAGR (Wilms' tumor, aniridia, genitourinary malformation, mental retardation), Drash syndrome (male pseudohermaphroditism and nephritis), and Perlman syndrome (visceromegaly, gigantism, cryptorchidism, polyhydramnios, classic facies), predispose to Wilms' tumor.[12–14] The risk of Wilms' tumor is 30% to 50% in children who have sporadic aniridia, 30% to 40% in children who have Drash syndrome, and 4% children who have Beckwith-Wiedemann syndrome.[12–14] Other genitourinary tract anomalies with increased incidence of Wilms' include renal ectopia, ureteral duplication, renal hypoplasia, cryptorchidism, and horseshoe kidney.[15]

Two loci on chromosome 11 also have been implicated in the genesis of Wilms' tumor.[10,12,13] Locus 11p13 locus, known as the WT1 gene, has been identified in patients with WAGR and Drash syndromes. Locus 11p1, known as the WT2 gene, has been linked to the Beckwith-Wiedemann syndrome or hemihypertrophy.[13] Evidence