Abdominal Masses

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Finding an abdominal mass on a child can be alarming to both the parents and pediatrician. Abdominal masses are often incidentally discovered by a parent while bathing the child, palpated unexpectedly on routine physical examination, or detected on abdominal imaging. The causes of pediatric abdominal masses are extensive, ranging from benign to neoplastic, and often originating from organs within the intra-abdominal cavity (Table).

At presentation, patients may be asymptomatic or report a wide range of associated symptoms, including fever, hematuria, and abdominal pain or distension. New-onset hypertension may be the first sign of an abdominal mass. The child's age, associated symptoms, location of mass, and laboratory findings provide important clues to the underlying cause and can direct appropriate evaluation and consultation.

Most abdominal masses in infants originate from the kidney and are benign. Often discovered prenatally, hydronephrosis, the most common renal mass in infants, is typically unilateral and results from vesicoureteral reflux and/or obstruction at the ureteropelvic junction. Multicystic dysplastic kidneys are another cause of hydronephrosis described as numerous noncommunicating cysts that vary in size, with little to no normal renal tissue identified. Bilateral renal masses detected on examination or imaging are concerning for autosomal recessive polycystic kidney disease (ARPKD). The presentation of ARPKD varies with the degree of hyperplasia of the collecting tubules and may be detected in utero or later in childhood.

The most common renal neoplasm in infants is congenital mesoblastic nephroma (CMN), which usually presents before 3 months of age as an asymptomatic abdominal mass but may be associated with hematuria, hypertension, hypercalcemia, and even congestive heart failure. Wilms tumor or nephroblastoma, the most common renal malignancy of childhood, primarily occurs between the ages of 2 and 5 years but can develop in infancy. Most cases are sporadic, although hereditary forms do exist. Patients may be asymptomatic or present with abdominal pain and hypertension. Unilateral involvement is most common. Children with WAGR (Wilms tumor, aniridia, genitourinary anomalies, retardation), Denys-Drash, and Beckwith-Wiedemann (BWS) syndromes are at increased risk for the development of Wilms tumor and require frequent screening. Clear cell sarcoma is the second most common childhood renal malignancy. An aggressive tumor typically found in children younger than age 4 years, it can present with hematuria, hypertension, and abdominal pain. During adolescence, renal cell carcinoma is the most common kidney tumor, classically presenting with painless gross hematuria, flank pain, and a palpable mass.

Liver masses account for 5% to 6% of all pediatric intra-abdominal masses and, unlike renal masses, are primarily malignant. Benign vascular tumors usually

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Pediatrics: Diagnosis of Neuroblastoma. Sharp SE, Gelfand MJ, Shulkin BL. Semin Nucl Med. 2011;41(5):345–353 affect the liver in infants. Hemangiomas are the most common and typically present before age 6 months. Often detected prenatally, congenital hemangiomas are usually solitary lesions that mature by birth and may involute by age 2 years or persist for life. Infantile hemangiomas of the liver are often associated with multiple hemangiomas of the skin. Initially these tumors grow rapidly, then involute, and typically resolve within the first decade of life. Kaposiform hemangioendothelioma is a rare vascular tumor that may present in infancy as a life-threatening consumptive coagulopathy known as Kasabach-Merritt syndrome.

Beyond infancy, hepatoblastoma is the most common primary liver tumor in children, often presenting between ages I and 3 years, although it can occur as late as adolescence. The tumor can be associated with genetic syndromes, such as BWS and Aicardi syndrome. Hepatoblastoma should be suspected in the child who has a large, solitary mass of the liver with elevated α -I-fetoprotein (AFP) concentrations. Hepatocellular carcinoma (HCC) can develop in children with liver disease, with cirrhosis serving as a significant risk factor. Children with a liver mass concerning for HCC often have vague symptoms of abdominal pain, weight loss, or decreased appetite and are usually between ages IO and I4 years.

Neuroblastoma is the most common infantile malignancy and most common extracranial solid tumor in children. Primarily affecting the adrenal glands, neuroblastoma typically presents before age 6 years (median age of 15 months) as a palpable abdominal mass with a wide spectrum of associated symptoms and a propensity to metastasize to bone marrow, cortical bone, the liver, and lymph nodes. Although uncommon, the paraneoplastic opsoclonusmyoclonus syndrome, known also as dancing eyes—dancing feet syndrome, can signal the occurrence of neuroblastoma.

Gastrointestinal (GI) duplication cysts can occur anywhere along the GI tract and are usually spherical cysts not connected with bowel lumen. GI tract duplications may serve as a lead point for a volvulus or intussusception. By far the most common cause of an abdominal mass in a preschool-age child around the time of toilet training is constipation, especially if irregular bowel movements, straining with defecation, or hard stools are reported. Constipation can be confirmed with disappearance of the mass after appropriate use of an osmotic agent.

Only rarely does an abdominal mass in an infant result from a primary tumor or cyst of the pancreas or spleen. Pancreatoblastoma does occur during the first decade of life, again very rarely, manifesting as either an asymptomatic abdominal mass or with vague GI complaints. An upper abdominal mass preceded by pancreatitis or trauma is likely a pancreatic pseudocyst. By enlarging the spleen, both Hodgkin and non-Hodgkin lymphoma can present as a

TABLE. Causes of Pediatric Abdominal Masses

Renal	
	Hydronephrosis
	Polycystic kidney disease
	Nephroblastoma (Wilms tumor)
	Congenital mesoblastic nephroma
	Clear cell carcinoma
	Anaplastic sarcoma
	Angiomyolipomas
	Nephroma
	Rhabdoid tumor
	Renal cell carcinoma
	Renal pseudotumor
	Renal vein thrombosis
	Metanephric stromal tumor
	Reninoma
Hepatobiliary	
	Hemangioma
	Hepatoblastoma
	Choledochal cyst
	Hepatocellular carcinoma
	Kaposiform hemangioendothelioma
Adrenal	
	Neuroblastoma
Gastrointestinal	
	Constipation
	Pyloric stenosis
	Intussusception
	Gastrointestinal duplication cyst

TABLE. (Continued)	
Pancreas	
	Pancreatic pseudocyst
	Pancreatoblastoma
Spleen	
	Hemangiomas
	Lymphangioma
	Hamartoma
	Hodgkin lymphoma
	Non-Hodgkin lymphoma
Genitourinary	
	Pregnancy
	Ovarian cyst
	Rhabdomyosarcoma
	Teratoma
	Germ cell tumor
	Urachal cyst

solitary abdominal mass. Other neoplastic tumors of the spleen are rarities. Although uncommon, benign vascular tumors can affect the spleen; they include hemangiomas, lymphangiomas, and hamartomas.

In older children and adolescents, masses can arise from the genitourinary tract. They can be benign (ovarian cysts, germ cell tumors, teratomas, pregnancy) or malignant (rhabdomyosarcomas).

Once an abdominal mass is identified, management usually consists of abdominal imaging to narrow the differential diagnosis. Abdominal radiographs help with determining location and origin and may detect calcium deposits, but ultrasonography is the preferred initial imaging modality and often the only imaging needed. Ultrasonography can identify the origin and consistency of the mass, along with evaluating the vascular supply with Doppler imaging. If a neoplastic tumor is suspected on ultrasonography or the imaging is unsuccessful, abdominal computed tomography scan or magnetic resonance imaging may be necessary to characterize the mass, assess organ involvement, and stage the disease. Metaiodobenzylguanidine (MIBG) scans are a

nuclear medicine study used to stage and evaluate response to therapy in neuroblastomas.

Initial laboratory evaluation typically includes a complete blood cell count, chemistry panel, liver function tests, and urinalysis. Polycythemia, normocytic anemia, pancytopenia, or marked leukocytosis may result from a malignancy. Hypercalcemia occurs with CMN and may also be a paraneoplastic phenomenon. Abnormal liver function test results may indicate a hepatobiliary origin. Urinalysis can indicate the presence of blood, suggesting a renal cause, especially if accompanied by proteinuria. Biomarkers may be helpful in establishing specific diagnoses, such as elevated AFP concentrations associated with both hepatoblastoma and pancreatoblastoma.

Treatment, prognosis, and outcomes of pediatric abdominal masses vary, based on the cause. Most pediatric masses are benign. Referral to a subspecialist or tertiary care center is warranted if the nature of the mass cannot be identified or if the mass requires subspecialty or surgical care.

COMMENT: In their discussion of abdominal masses arising within the GI tract, Drs Potisek and Antoon left out one I find particularly interesting. Bezoars are accumulations of undigested material, most often occurring in the stomach, that can enlarge enough to cause signs and symptoms of obstruction and become palpable as epigastric masses. Two forms of bezoars are especially relevant to pediatricians: lactobezoars in neonates, especially preterm infants, and infants; and trichobezoars in older children and adolescents.

Lactobezoars are gastric concretions of milk (or formula) and mucus that can lead to poor feeding, distention, irritability, vomiting, and poor weight gain, symptoms that are not very different from the more common pyloric stenosis. Gastric trichobezoars, matted collections of hair in the stomach, are often the product of trichotillomania – compulsive hair-pulling that may be accompanied by anxiety and/or depression. They too can lead to abdominal distention and pain, vomiting, weight loss, and even bleeding and perforation.

If we don't think of them...

Henry M. Adam, MD
 Associate Editor, In Brief

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