Pediatric Hepatobiliary Neoplasms
An Overview and Update

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KEYWORDS

• Children • Neoplasms • Liver • Radiology • Radiography • Ultrasonography • Computed tomography • MR Imaging

KEY POINTS

- The most commonly encountered malignant tumors of the liver are hepatoblastoma in infants and young children and fibrolamellar carcinoma and hepatocellular carcinoma in older children.
- Infantile hepatic hemangioma is the most commonly encountered liver tumor with typical imaging findings in infants.
- Serum alpha-fetoprotein level is typically increased in children with hepatoblastoma and hepatocellular carcinoma.
- Rhabdomyosarcoma is the main malignant tumor involving the biliary tree in the pediatric population, and often presents as an intraductal mass with a grape-like or branching pattern.
- The most common site of origin of liver metastases in children with solid tumors is neuroblastoma, followed by Wilms tumor.

INTRODUCTION

Hepatobiliary neoplasms constitute a wide range of liver tumors in children (**Box 1**). Primary hepatic neoplasms account for 1% to 4% of all solid tumors of childhood with approximately two-thirds of the primary hepatic neoplasms being malignant (**Fig. 1**). The incidence rate of malignant primary hepatic neoplasms is 1.5 per 1 million annually, resulting in 100 to 150 new cases every year in the United States.[1–3] The most common cause of metastatic disease of the liver is neuroblastoma, followed by Wilms tumor.[4] Rhabdomyosarcoma arising from the biliary tree is rare, accounting for approximately 1% of all rhabdomyosarcomas in childhood. The age of the patient, underlying hepatic disease, and laboratory tests are helpful when evaluating the imaging studies of children with hepatobiliary tumors (**Table 1**).

Recent advances in chemotherapy options, surgical approaches and transplant options for unresectable tumors, and collaboration between international pediatric oncology groups have refined the management of hepatobiliary neoplasm in pediatric patients. Accurate radiologic interpretation, correlated with supportive demographic data and laboratory findings, is critical to the diagnosis and proper treatment of hepatobiliary tumors in the pediatric population. This review article provides an overview and update on imaging strategy and characteristic imaging findings of hepatobiliary neoplasms in infants and children.

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Box 1
Common hepatobiliary neoplasms in children

Primary neoplasms of the liver

Infants and young children

Benign tumors

- Infantile hepatic hemangioma
- Mesenchymal hamartoma

Malignant tumors

- Hepatoblastoma
- Malignant rhabdoid tumor
- Undifferentiated embryonal sarcoma
- Angiosarcoma

Older children

Benign tumors

- Focal nodular hyperplasia
- Adenoma
- Nodular regenerative hyperplasia

Malignant tumors

- Hepatocellular carcinoma
- Fibrolamellar carcinoma
- Epithelioid hemangioendothelioma

Metastatic neoplasms of the liver

- Neuroblastoma
- Wilms tumor
- Lymphoma

Primary neoplasm of the biliary tree

- Rhabdomyosarcoma

IMAGING EVALUATION STRATEGY

Various non-invasive imaging modalities are currently available to image tumors of the liver and biliary tract in children. Each imaging modality provides complementary information, which, when taken as a whole, allows the radiologist to diagnose and stage the tumor and determine the most effective course of management.

Ultrasonography (US) is usually the first imaging modality used to exclude a hepatobiliary mass in the pediatric population. If there is no mass in the liver on US, no further imaging is necessary. If a mass is found, then magnetic resonance (MR) imaging of the abdomen should be subsequently performed. Computed tomography (CT) of the abdomen should only be considered if MR imaging is not available or contraindicated. If the characterization of the mass yields a benign lesion, the stability of the lesion can be monitored by US or MR imaging. If the CT or MR imaging yields a malignant mass, CT of the chest should be performed to evaluate for the presence of metastatic lung disease. A biopsy of the primary liver tumor should then be completed in order to determine histopathologic diagnosis.[2]

IMAGING MODALITY AND TECHNIQUE

Radiography

Radiographs may provide useful information by providing a global view of the abdomen and excluding other potential causes for the patients' symptoms. They may demonstrate the extent of the large hepatobiliary tumors and their mass effect on adjacent intra-abdominal structures, guiding the investigation toward the organ of interest or additional modalities. Calcifications, when present

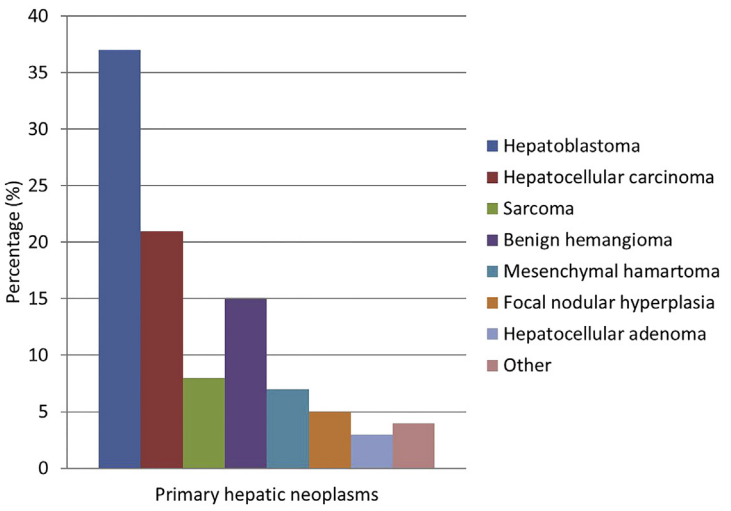

Fig. 1. The chart illustrates the frequency of common primary hepatic neoplasms in children. (*Data from* Rozell JM, Catanzano T, Polansky SM, et al. Primary liver tumors in pediatric patients: proper imaging technique for diagnosis and staging. Semin Ultrasound CT MR 2014;35:382–93.)