Pediatric Liver Tumors

Kenneth Ng, DO, Douglas B. Mogul, MD, MPH*

KEYWORDS

• Liver • Tumor • Pediatrics • Evaluation • Diagnosis • Management

KEY POINTS

- Although liver tumors are rare in the pediatric population, they are common in the setting of children with specific risk factors requiring increased awareness and, in some instances, screening.
- The evaluation of a liver mass in children is largely driven by the age at diagnosis, the presence of any medical comorbidities, and initial testing with alpha fetoprotein and imaging.
- Specific guidelines for the management of different tumors have been implemented in recent years such that a multidisciplinary approach is ideal and care should be provided by centers with experience in their management.

INTRODUCTION

Liver tumors in childhood are rare. Approximately two-thirds of all pediatric liver tumors are malignant, including hepatoblastoma (HB; 37%), hepatocellular carcinoma (HCC; 21%), and sarcoma (8%), and these cancers comprise approximately 1% of all childhood tumors reported to the Surveillance, Epidemiology, and End Results (SEER) registry.[1–3] Benign tumors (vascular lesions such as hemangiomas and hemangioendotheliomas [15%], focal nodular hyperplasia [5%], mesenchymal hamartomas [7%]) can also be clinically challenging if they are large enough to compress neighboring organs and replace normal-functioning hepatic tissue.

Although uncommon, it is important that general pediatricians as well as subspecialists in pediatric gastroenterology and hepatology continue to be aware of the epidemiology, clinical presentation, and the initial approach to the diagnosis of these diseases. First, these tumors may be more frequently seen in specific subgroups, such as in individuals with genetic conditions associated with increased cancer risk as well as children with specific risk factors such as prematurity (**Table 1**).[4,5] Furthermore, evidence exists that the incidence of HB is increasing, with a doubling of the rate from 0.6

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Department of Pediatrics, Johns Hopkins University School of Medicine, 600 North Wolfe Street, CMSC 2-117, Baltimore, MD 21287, USA
* Corresponding author.
E-mail address: dmogul1@jhmi.edu

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Table 1
Risk factors for hepatoblastoma and hepatocellular carcinoma in children

Risk Factor	HB	HCC
Extreme prematurity and low birthweight	X	—
Total parenteral nutrition	—	X
Infections		
Hepatitis B virus	—	X
Hepatitis C virus	—	X
Tumor syndromes		
Familial adenomatous polyposis and Gardner syndrome	X	X
Beckwith-Wiedemann	X	—
Neurofibromatosis	—	X
Li-Fraumeni	X	—
Glycogen storage disease (type I and III)	X	X
Hereditary tyrosinemia	—	X
Congenital cholestasis		
Biliary atresia	—	X
Progressive familial intrahepatic cholestasis type 2	—	X
Alagille syndrome	—	X
α-1-antitrypsin deficiency	—	X
Other Genetic Diseases		
Trisomy 21	X	—
Ataxia telangiectasia	—	X
Fanconi anemia	—	X
Medications		
Methotrexate	—	X
Oral contraceptives	—	X
Anabolic steroids	—	X
Underlying Liver Disease		
Congenital hepatic fibrosis	—	X
Congenital portosystemic shunt	X	X
Hepatic adenoma	—	X

cases per million between 1973 and 1977 to 1.2 cases per million between 1993 and 1997, and this can likely be attributed to the increased survival of premature infants.[1,6] Third, the increasing use of computed tomography (CT) and ultrasonography scanning for the routine evaluation of children in the emergency room and other settings has led to a corresponding increase in the identification of incidental findings of hepatic lesions in children.[7] Pediatricians may therefore see increasing numbers of these masses over time.

CLINICAL PRESENTATION

Most liver masses are asymptomatic and are detected through palpation by either a parent or a physician.[5,8] In rare instances, children present with complications stemming from obstruction of major bile ducts by the mass, leading to jaundice and pruritus, as well as fever (ie, cholangitis) and biliary colic. Compression of hepatic