

- In patients with pregnancy plans or LT candidates, elective interventions in patients with HAAs or SAAs should be considered. In other patients, intervention has not been proven superior to expectant management.

Issues Specific to Children and Congenital Disorders

EXTRAHEPATIC PORTAL VEIN OBSTRUCTION

Extrahepatic portal vein obstruction (EHPVO) has also been referred to as cavernous transformation of the PV in children. It is believed to be an acquired lesion subsequent to a number of postnatal events, including umbilical vein catheterization, omphalitis, and *de novo* thrombosis, attributable to hypercoagulable conditions. However, the embryology of the PV formation is complex, and at any one of a number of steps in PV formation, the final integrity of the PV may be compromised during development. The clinical hallmarks of EHPVO, as with any form of portal hypertension, are primarily those of hemorrhage from gastric and esophageal varices and hypersplenism with splenic enlargement, thrombocytopenia, and, less commonly, ascites. Liver function is always preserved unless EHPVO occurs with an accompanying coexistent primary liver disorder. Evidence suggests that early intervention with restoration of portal blood flow confers an advantage, and delayed reconstruction may be detrimental to the final outcome.⁽³¹⁰⁾ Treatment consists of management of the complications of portal hypertensive bleeding and consultation for possible shunt surgery. The meso-Rex bypass (surgical extrahilar mesenterico/left portal shunt⁽³¹¹⁾) has been established as the primary and preferred surgical correction for EHPVO, but portosystemic shunting with either a distal splenorenal shunt or a mesocaval shunt is indicated if the meso-Rex bypass is anatomically not possible.⁽³¹²⁾ The meso-Rex bypass corrects some of the physiological abnormalities subsequent to chronic portosystemic shunting, such as minimal hepatic encephalopathy and coagulation abnormalities.⁽³¹³⁻³¹⁵⁾ Given the advancing confidence in the results of the meso-Rex bypass, it

has been proposed that corrective surgery should be considered in asymptomatic children to prevent the development of portal hypertension complications and downstream sequelae.⁽³¹⁶⁾ Evaluation and surgical treatment of these children should be in hospitals with expertise in hepatobiliary surgical care and transplantation in children.⁽³¹⁶⁾

Hepatic Hemangiomas

In recent years, there have been a number of changes in classification and treatment recommendations of hepatic hemangiomas in children.⁽³¹⁷⁾ These may be classified into two types: (1) congenital (rapidly involuting congenital hemangioma and noninvoluting congenital hemangioma) and (2) infantile (focal, diffuse, and multifocal). In general, the distinction between the congenital and infantile types of hemangiomas is blurred. Symptoms can range from none to cardiac decompensation attributable to high-output heart failure. The focal type rarely causes symptoms, and most can safely be observed. Many involute or remain stable. Hemangioma histopathological glutathione expression may occur in as many as 30%-50% of cases of infantile hemangiomata,⁽³¹⁸⁾ and these have been shown to respond to nonselective beta-blockers such as propranolol.^(319,320) The multifocal or diffuse types may require interventions, ranging from medical therapies, such as beta-blockers or embolization, to surgical resection or LT in the most extreme examples. Other treatment depends primarily on symptoms and includes observation for many, medical therapies for heart-related symptoms, embolization, or hepatic arterial ligation as a means to reduce cardiac output, surgical resection for local or lobar disease, and LT primarily for those not responding to therapy or unresectable symptomatic cases.

Hepatic Arteriovenous Malformations

Hepatic arteriovenous malformations are rare lesions that can be acquired or congenital. Acquired lesions are generally secondary to external trauma, such as a penetrating injury to the liver. Iatrogenic injuries may also be possible secondary to percutaneous liver