

Background and Purpose

Background:

Interventional Radiologists are commonly consulted for evaluation and management of pediatric patients with underlying portal hypertension. There are key differences between this disease process in adults and pediatric patients that alters the natural history of disease, workup, and management of the patients.

Purpose:

- Understand the common etiologies of portal hypertension in children and how they contrast with those affecting adults
- Discuss different methods of anatomic liver transplantation in children, and how this affects treatment of liver disease in children
- Recognize the primary role of endoscopic surveillance and therapy for treatment of variceal bleeding in children
- Understand surgical shunting approaches in pediatric portal hypertension patients

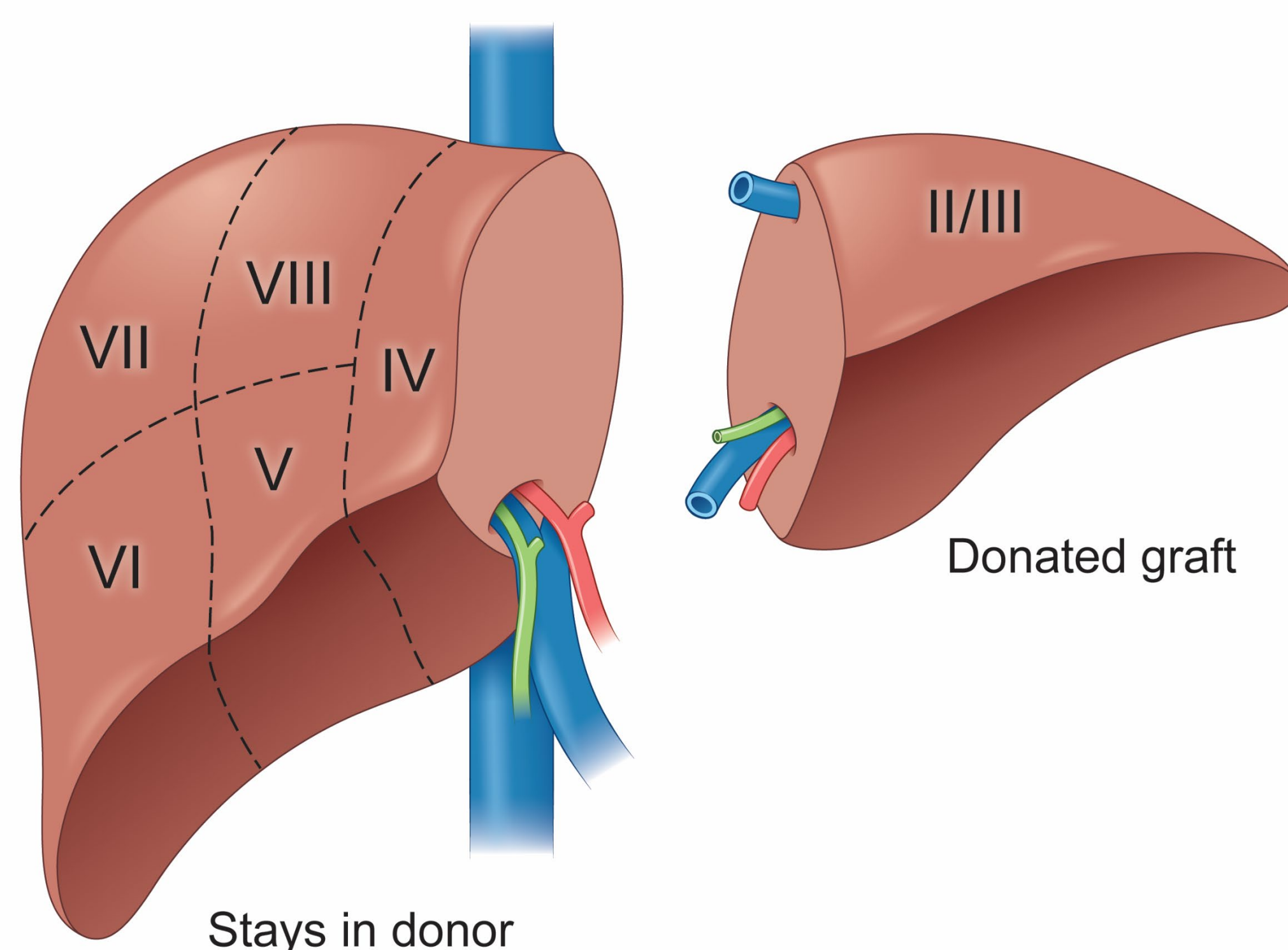
Discussion

The underlying causes of portal hypertension in children are significantly different than those seen in adults. Extrahepatic portal vein obstruction (EHPVO) is one of the most common etiologies of portal hypertension seen in children. Patients with EHPVO have been linked to an increased risk of developing esophageal varices and hypersplenism when compared to other causes of portal hypertension.

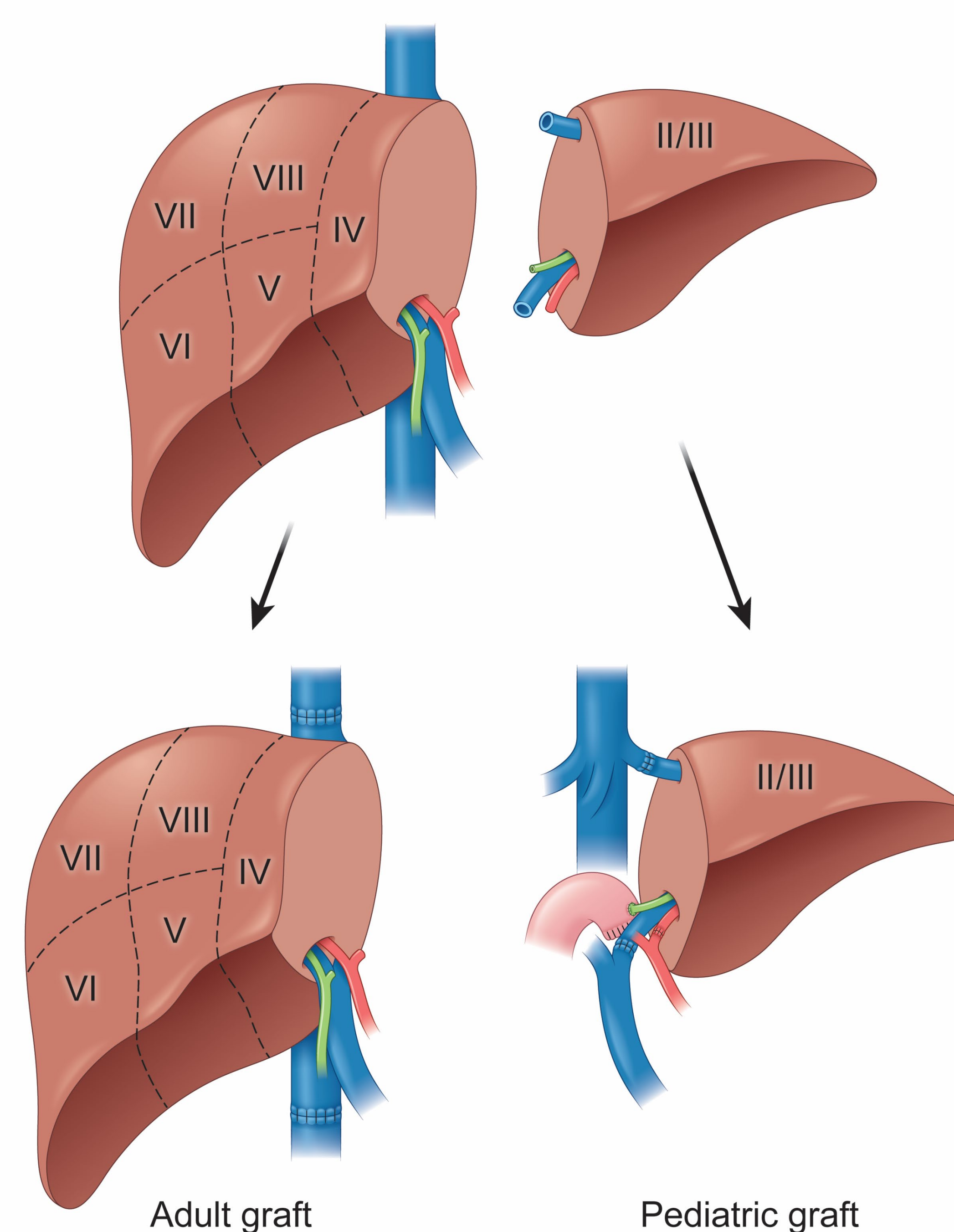
Liver transplantation is often the definitive treatment for cirrhosis with portal hypertension in children, and graft supply for children is relatively high when compared to adults. This is commonly pursued in cases of portal hypertension due to cirrhosis, such as in biliary atresia. In other patients, conventional medical and endoscopic management is relied upon.

Illustrations

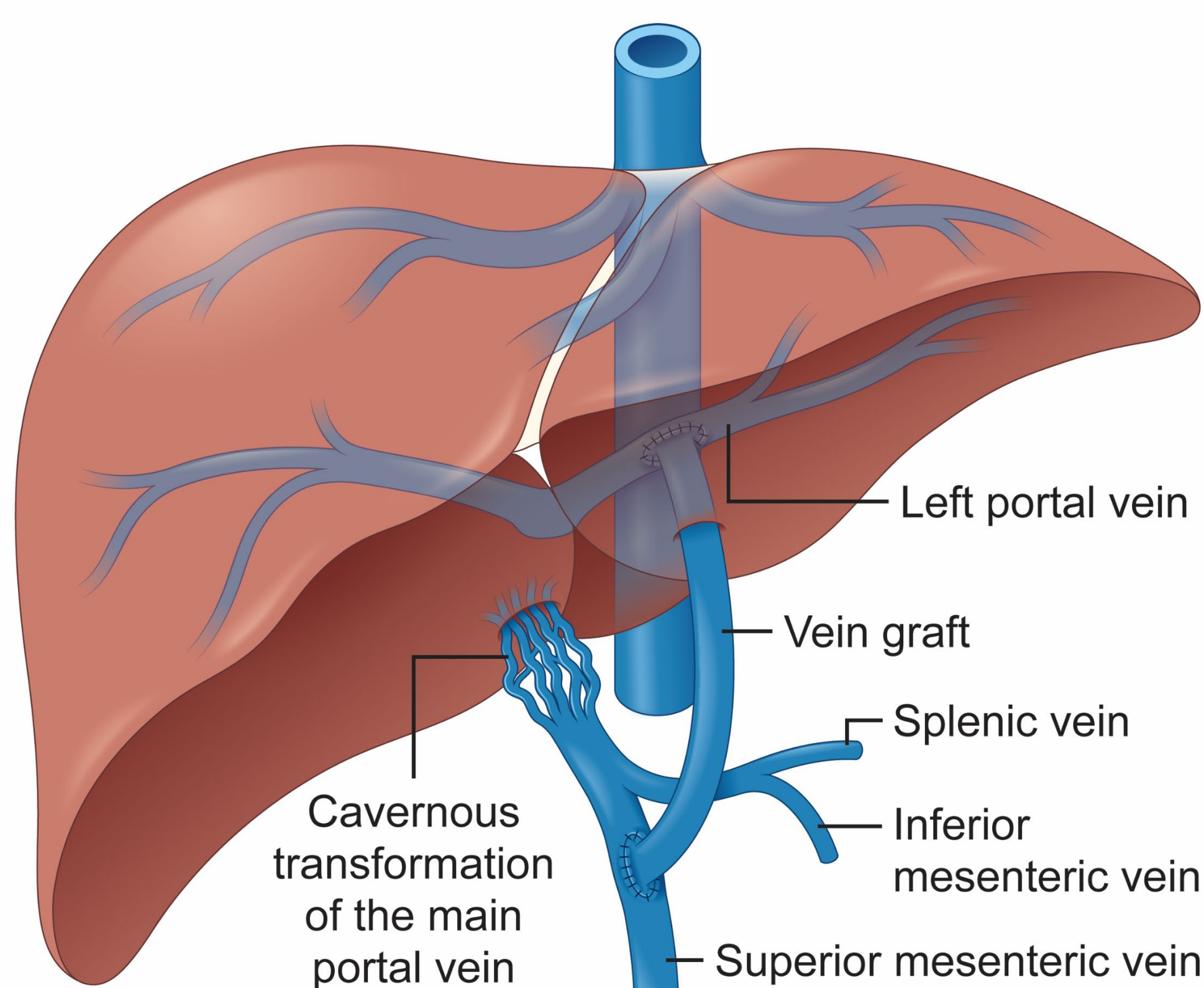
Left Lateral Segment Transplantation Anatomy



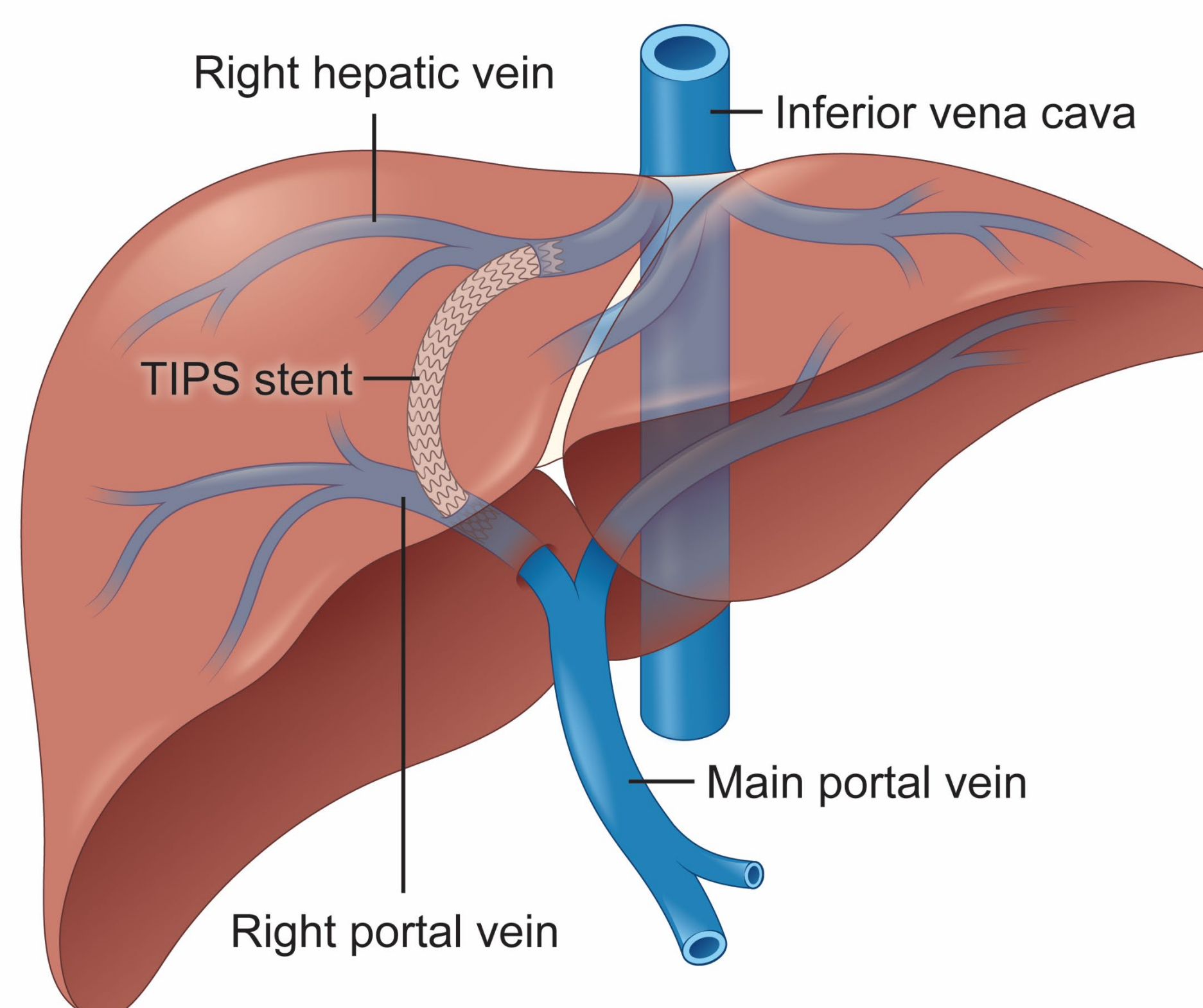
Split Liver Transplantation Anatomy



Meso-Rex Bypass Anatomy



TIPS Procedure Anatomy



Discussion Continued

Endoscopy remains the primary treatment modality for bleeding esophageal varices in children due to portal hypertension, though data on which patients with suspected portal hypertension should undergo endoscopic surveillance is a topic of ongoing research. Endoscopic banding is generally superior to endoscopic sclerotherapy, and sclerotherapy is generally reserved for patients in whom band ligation is not possible due to their small size.

Surgical shunting in the form of mesenteric vein bypass remains popular in pediatric patients, partly due to anatomic considerations in children with isolated extrahepatic portal vein thrombosis. The Meso-Rex bypass allows for restoration of near-anatomic flow via an autologous shunt from the distal SMV to the intrahepatic portal venous system, negating risks of encephalopathy and CHF associated with more shunts to the hepatic veins such as TIPS.

TIPS in children for palliation of severe symptoms of portal hypertension and as second line therapy for bleeding esophageal varices among other indications is performed. However, heightened concerns over the negative effects of hepatic encephalopathy and long-term shunt durability in children makes TIPS less popular in this population.

Conclusions

An IR working with children with portal hypertension must understand the key differences in etiology and treatment amongst this patient population. TIPS may still be pursued, but has different implications in children.

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