INTRODUCTION

The survival of children with portal hypertension has improved over the past decade owing to significant improvements in both medical and surgical therapy. These improvements have included progress in the pharmacologic control of acute portal hypertensive hemorrhage and improved efficacy and safety of endoscopic methods to treat acute esophageal variceal hemorrhage. However, despite these changes, there remains a significant role for advanced surgical therapy using portocaval shunts in children with gastrointestinal
hemorrhage were significant sequela of portal hypertension. In addition, patience with both irreversible liver disease and portal hypertension have benefited from the improved success with pediatric liver transplantation as a definitive treatment for children with end-stage liver disease. This review will primarily examine the role of surgical therapy for children with progressive portal hypertension.

DEFINITION, ETIOLOGY

Portal hypertension is defined as an elevation of the portal pressure above 10–12 mmHg. In healthy children, portal pressure rarely exceeds 7 mmHg. Elevation of the portal pressure is commonly classified by the anatomic location of the obstructed portal venous flow, subdivided as presinusoidal, sinusoidal, or postsinusoidal block, although increased splanchnic blood flow may contribute in some cases. The response to increased portal venous pressure is similar to adults, with the development of collateral circulatory pathways connecting the high-pressure portal vasculature to the low-pressure systemic venous system. The most common communications occur within the esophageal wall, connecting the coronary and short gastric veins to the esophageal venous plexus, which communicate with the intercostal,azygous, and hemiazzygous veins. As portal pressure increases, esophageal varices developing within this plexus become the site with the highest risk for massive hemorrhage. Less-threatening collateral communications can develop between the recanalized umbilical vein and abdominal wall systemic veins (caput medusa), the inferior rectal veins as hemorrhoids, and around the retroperitoneal pancreas and duodenum. In addition, any surgical union between the portal and systemic venous circulation, such as occurs with intestinal stomas or previous incision sites, can become problematic collateral sites. Favorable collaterals developing within the tissues surrounding the pancreas, duodenum, and left kidney form “spontaneous” spleno-renal shunts. The possibility that these collaterals play a significant role in ultimately decreasing portal venous pressure and preventing variceal hemorrhage has been suggested, but remains unproven. In our opinion, their radiographic and physical appearance exceeds their hemodynamic importance and benefit.

The progressive development of collaterals connecting the portal and systemic circulation has the theoretical beneficial effect of decreasing portal pressure. However, this effect is ameliorated by the concurrent development of a hyperdynamic circulatory state (1). Portal hypertension has been associated with the presence of autonomic nervous system dysfunction, and an excess of circulating cytokines leading to tachycardia, decreased systemic, and splanchnic vascular resistance secondary to vasodilatation, plasma volume expansion, increased cardiac output, and subsequently, increased portal inflow.

The combination of increasing portal inflow, venous outflow obstruction, and the remarkable collateral circulation that develops account for many of the complications associated with portal hypertension. Superficial submucosal varices, especially those in the esophagus and stomach, and, to a lesser extent, those in the duodenum, colon, or rectum, are prone to rupture and bleeding. In addition, prominent submucosal arteriovenous communications between the muscularis mucosa and dilated precapillaries and veins within the stomach result in vascular ectasia, or congestive hypertensive gastropathy, significantly contributing to the risk of hemorrhage from the stomach.

Each of the causes of elevated portal pressure shares the common mechanism of increased resistance to blood flow from the visceral/splanchnic portal circulation to the right atrium.