Preface: Portal Hypertension

Sammy Saab

Portal Hypertension: Pathogenesis and Diagnosis

Laura Turco and Guadalupe Garcia-Tsao

Portal hypertension (PH) is an increase in the pressure gradient between portal vein and inferior vena cava. Increased resistance occurs at different levels within the portal venous system, followed by increased portal venous inflow. PH is the main driver of cirrhosis decompensation. Varices on endoscopy or portosystemic collaterals on imaging indicate PH. Although its cause is determined mostly via noninvasive tests, the gold standard to measure portal pressure in cirrhosis and determine its severity is hepatic vein catheterization with determination of the hepatic venous pressure gradient. Measuring portal pressure is essential in proof-of-concept studies of portal pressure-lowering drugs.

Frailty, Sarcopenia, and Malnutrition in Cirrhotic Patients

Elizabeth S. Aby and Sammy Saab

Sarcopenia, frailty, and malnutrition are prevalent complications in patients with end-stage liver disease (ESLD) and are associated with increased risk of morbidity and mortality. It is valuable to measure nutritional status, sarcopenia, and frailty over time in order to create interventions tailored to individuals with ESLD. Evaluating sarcopenia and frailty in patients with ESLD is challenging. Further work is needed to perfect these assessments so that clinicians can incorporate these assessments into their decision-making and management plans for cirrhotic patients.

Hepatic Encephalopathy Challenges, Burden, and Diagnostic and Therapeutic Approach

Beshoy Yanny, Adam Winters, Sandra Boutros, and Sammy Saab

Hepatic encephalopathy (HE) is an important cause of morbidity and mortality in patients with cirrhosis. The impact of HE on the health care system is similarly profound. The number of hospital admissions for HE has increased in the last 10-year period. HE is a huge burden to the patients, care givers, and the health care system. HE represents a “revolving door” with readmission, severely affects care givers, and has effects on cognition that can persist after liver transplant. This article reviews the current literature to discuss the challenges and diagnostic and therapeutic approaches to HE.

Varices: Esophageal, Gastric, and Rectal

Thomas O.G. Kovacs and Dennis M. Jensen

Gastrointestinal varices are associated with cirrhosis and portal hypertension. Variceal hemorrhage is a substantial cause of morbidity and
mortality, with esophageal and gastric varices the most common source and rectal varices a much less common cause of severe gastrointestinal bleeding. The goals of managing variceal hemorrhage are control of active bleeding and prevention of rebleeding. This article focuses on reviewing the current management strategies, including optimal medical, endoscopic, and angiographic interventions and their clinical outcomes to achieve these goals. Evidence based discussion is used with current references as much as possible.

An Update: Portal Hypertensive Gastropathy and Colopathy

Don C. Rockey

Complications of portal hypertension include portal hypertensive gastropathy and colopathy. These disorders may cause chronic or acute gastrointestinal bleeding. The diagnosis is made endoscopically; therefore, there is great variability in their assessment. Portal hypertensive gastropathy can range from a mosaic-like pattern resembling snakeskin mucosa to frankly bleeding petechial lesions. Portal hypertensive colopathy has been less well-described and is variably characterized (erythema, vascular lesions, petechiae). Treatment is challenging and results are inconsistent. Currently, available evidence does not support the use of beta-blockers for primary prevention. Further investigation of the pathogenesis, natural history, and treatment of these disorders is needed.

Ascites and Hepatorenal Syndrome

Danielle Adebayo, Shuet Fong Neong, and Florence Wong

Ascites occurs in up to 70% of patients during the natural history of cirrhosis. Management of uncomplicated ascites includes sodium restriction and diuretic therapy, whereas that for refractory ascites (RA) is regular large-volume paracentesis with transjugular intrahepatic portosystemic shunt being offered in appropriate patients. Renal impairment occurs in up to 50% of patients with RA with type 1 hepatorenal syndrome (HRS) being most severe. Liver transplant remains the definitive treatment of eligible candidates with HRS, whereas combined liver and kidney transplant should be considered in patients requiring dialysis for more than 4 to 6 weeks or those with underlying chronic kidney disease.

Pulmonary Complications of Portal Hypertension

Rodrigo Cartin-Ceba and Michael J. Krowka

The most common pulmonary complications of chronic liver disease are hepatic hydrothorax, hepatopulmonary syndrome, and portopulmonary hypertension. Hepatic hydrothorax is a transudative pleural effusion in a patient with cirrhosis and no evidence of underlying cardiopulmonary disease. Hepatic hydrothorax develops owing to the movement of ascitic fluid into the pleural space. Hepatopulmonary syndrome and portopulmonary hypertension are pathologically linked by the presence of portal hypertension; however, their pathophysiologic mechanisms are significantly different. Hepatopulmonary syndrome is characterized by low pulmonary vascular resistance secondary to intrapulmonary vascular dilatations and hypoxemia; portopulmonary hypertension features elevated pulmonary
vascular resistance and constriction/obstruction within the pulmonary vasculature.

Pharmacologic Management of Portal Hypertension
Chalermrat Bunchorntavakul and K. Rajender Reddy

Terlipressin, somatostatin, or octreotide are recommended as pharmacologic treatment of acute variceal hemorrhage. Nonselective β-blockers decrease the risk of variceal hemorrhage and hepatic decompensation, particularly in those 30% to 40% of patients with good hemodynamic response. Carvedilol, statins, and anticoagulants are promising agents in the management of portal hypertension. Recent advances in the pharmacologic treatment of portal hypertension have mainly focused on modifying an increased intrahepatic resistance through nitric oxide and/or modulation of vasoactive substances. Several novel pharmacologic agents for portal hypertension are being evaluated in humans.

Role of Transjugular Intrahepatic Portosystemic Shunt in the Management of Portal Hypertension: Review and Update of the Literature
Matthew L. Hung and Edward Wolfgang Lee

Transjugular intrahepatic portosystemic shunt (TIPS) is a well-established procedure used in the management of complications of portal hypertension. Although the most robust evidence supports the use of TIPS as salvage therapy in variceal hemorrhage, secondary prophylaxis of variceal bleeding, and treatment of refractory ascites, there is also data to suggest its efficacy in other indications such as hepatic hydrothorax, hepatorenal syndrome, and Budd-Chiari syndrome. Recent literature also suggests that TIPS may improve survival for certain subpopulations if placed early after variceal bleeding. This article provides an updated evidence-based review of the indications for TIPS. Outcomes, complications, and adequate patient selection are also discussed.

Surgery in Patients with Portal Hypertension
Melissa Wong and Ronald W. Busuttil

Patients with portal hypertension will increasingly present for nontransplant surgery because of the increasing incidence of, and improving long-term survival for, chronic liver disease. Such patients have increased perioperative morbidity and mortality caused by the systemic pathophysiology of liver disease. Preoperative assessment should identify modifiable causes of liver injury and distinguish between compensated and decompensated cirrhosis. Risk stratification, which is crucial to preparing patients and their families for surgery, relies on scores such as Child-Turcotte-Pugh and Model for End-stage Liver Disease to translate disease severity into quantified outcomes predictions. Risk factors for postoperative complications should also be recognized.

Noncirrhotic Portal Hypertension: Current and Emerging Perspectives
Rajeev Khanna and Shiv Kumar Sarin

Idiopathic portal hypertension (IPH) and extrahepatic portal venous obstruction (EHPVO) are prototype noncirrhotic causes of portal
hypertension (PHT), characterized by normal hepatic venous pressure gradient, variceal bleeds, and moderate to massive splenomegaly with preserved liver synthetic functions. Infections, toxins, and immunologic, prothrombotic and genetic disorders are possible causes in IPH, whereas prothrombotic and local factors around the portal vein lead to EHPVO. Growth failure, portal biliopathy, and minimal hepatic encephalopathy are long-term concerns in EHPVO. Surgical shunts and transjugular intrahepatic portosystemic shunt resolve the complications secondary to PHT. Meso-Rex shunt is now the standard-of-care surgery in children with EHPVO.