PORTAL HYPERTENSION

Tianjin Medical University
LIU JIAN
DEFINITION

- Portal hypertension is present if portal venous pressure exceeds 10mmHg (1.3kPa).
- Normal portal venous pressure is 5–10mmHg (0.7–1.3kPa), which is sufficient to maintain a portal flow through the hepatic sinusoids of approximately 1 liter/min.
CLASSIFICATION AND CAUSES

Presinusoidal

- Thrombosis of the extrahepatic portal vein or one of its major tributaries—extrahepatic
- Pathologic processes that affect the terminal portal venules in the presinusoidal position—intrahepatic
- Liver function is well preserved
- Variceal bleeding
Sinusoidal

- caused by cirrhosis, and viral hepatitis and ethanol abuse, autoimmune liver disease, hemochromatosis
- Hepatocellular damage, destroyed sinusoidal anatomy, regenerative nodules form, fibrosis further obstructs portal venous flow and the normal metabolic processes are disrupted.
- Evaluate liver function, assess the activity of the underlying liver disease
Postsinusoidal

- Results from hepatic venous outflow obstruction—— Budd–Chiari syndrome.
- The least common cause of portal hypertension, but may be reversible if identified and treated early
**Etiologies of portal hypertension.** These are divided into those associated with normal liver function (presinusoidal) and those associated with liver damage (sinusoidal).

<table>
<thead>
<tr>
<th>Portal vein</th>
<th>Sinusoids</th>
<th>Hepatic veins</th>
</tr>
</thead>
<tbody>
<tr>
<td>Extrahepatic</td>
<td>Intrahepatic</td>
<td></td>
</tr>
<tr>
<td>Congenital</td>
<td>Schistosomiasis</td>
<td>Budd–Chiari syndrome</td>
</tr>
<tr>
<td>Umbilical sepsis</td>
<td>Congenital hepatic fibrosis</td>
<td>Veno-occlusive disease</td>
</tr>
<tr>
<td>Trauma</td>
<td>Primary biliary cirrhosis</td>
<td></td>
</tr>
<tr>
<td>Hypercoagulation state</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Malignant occlusion</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Portal venous anatomy in portal hypertension. The superior mesenteric, splenic and inferior mesenteric veins unite behind the neck of the pancreas to form the portal vein. The right and left gastric and gastroepiploic veins enter the portal and superior mesenteric veins.
### PATHOPHYSIOLOGY

#### STAGES IN THE DEVELOPMENT OF PORTAL HYPERTENSION

- Increased resistance to portal venous flow
- Formation of portal systemic collaterals
- Dilatation of the splanchnic venous bed and increased splanchnic flow
- Expansion of the intravascular plasma volume
- Peripheral and splanchnic vasodilatation leading to development of a hyperkinetic systemic circulation
HEMODYNAMIC PATHOPHYSIOLOGIC CHANGES

- Portal venous pressure increases to 20mmHg or greater, and blood is transmitted back into the whole splanchnic bed. The major sites for collateral pathway development are the natural watersheds for portosystemic collaterals: Gastroesophageal; Hemorrhoidal; Periumbilical; Retroperitoneal.
- Increased portal venous pressure, dilatation of the splanchnic venous bed and the development of collaterals all act as stimuli for splanchnic hyperemia.
- The clinical consequences are that a hyperdynaminc systemic circulation develops with a high cardiac output and a low total systemic vascular resistance and a low normal blood pressure.
Portal-Systemic Anastomosis
At the lower third of the esophagus, the esophageal branches of the left gastric vein (portal tributary) anastomose with the esophageal veins draining the middle third of the esophagus into the azygos veins (systemic tributary).
Half way down the anal canal, the **superior rectal veins** (portal tributary) draining the upper half of the anal canal anastomose with the **middle and inferior rectal veins** (systemic tributaries).
• The **paraumbilical veins** connect the left branch of the portal vein with the superficial veins of the anterior abdominal wall (systemic tributaries). The paraumbilical veins travel in the falciform ligament and accompany the ligamentum teres.
The veins of the ascending colon, descending colon, duodenum, pancreas, and liver (portal tributaries) anastomose with the renal, lumbar, and phrenic veins (systemic tributaries).
**CLINICAL PRESENTATION**

### PRESENTING FEATURES OF PORTAL HYPERTENSION

- Variceal bleeding
- Ascites
- Encephalopathy
- Splenomegaly, hypersplenism
- Incidental diagnosis
Classification of the severity of the underlying liver disease at the time of presentation of a patient who has portal hypertension is important. Child’s classification has stood the test of time.

Child’s original classification used the three clinical parameters of ascites, encephalopathy and nutrition plus the two biochemical parameters of bilirubin and albumin with a grading of 1–3 for each.

The Child’s classification modification most commonly used is that of Pugh, in which nutritional status is replaced by prothrombin time.
### Child–Pugh classification

#### ALLOCATION OF POINTS USING THE CHILD–PUGH CLASSIFICATION

<table>
<thead>
<tr>
<th>Parameter</th>
<th>1 point</th>
<th>2 points</th>
<th>3 points</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bilirubin (mg/dl)</td>
<td>&lt;2</td>
<td>2–3</td>
<td>&gt;3</td>
</tr>
<tr>
<td>Albumin (g/dl)</td>
<td>&gt;3.5</td>
<td>2.8–3.5</td>
<td>&lt;2.8</td>
</tr>
<tr>
<td>Increase in prothrombin time (s)</td>
<td>1–3</td>
<td>4–6</td>
<td>&gt;6</td>
</tr>
<tr>
<td>Ascites</td>
<td>None</td>
<td>Slight</td>
<td>Moderate</td>
</tr>
<tr>
<td>Encephalopathy</td>
<td>None</td>
<td>1–2</td>
<td>3–4</td>
</tr>
</tbody>
</table>

**Fig.27-6.** Child–Pugh classification. The grades are A for 5–6 points, B for 7–9 points and C for 10–15 points.
INVESTIGATION

HISTORY

• Exposure to hepatitis, a history of excessive alcohol ingestion or a family history of liver disease are relevant in history taking.

• Any history of upper gastrointestinal bleeding should be clarified in detail, with particular emphasis on the severity, occurrence of multiple episodes and management modalities used.
PHYSICAL EXAMINATION

• General physical examination should include a visual inspection for muscle wasting, jaundice and spider angiomas, and in males for gynecomastia and testicular atrophy.

• Abdominal examination focuses on hepatosplenomegaly and ascites, but may also reveal prominent abdominal wall veins.

• Systemic hemodynamic changes, which are manifested as a mild persistent tachycardia and a low blood pressure.
HEMATOLOGY

- Hematocrit is often reduced, either secondary to chronic bleeding or as a result of the expanded plasma volume;
- Platelet count may be less than $100 \times 10^9/\text{mm}^3$ and the white cell count less than $4 \times 10^9/\text{mm}^3$ because of hypersplenism.
- Prolongation of the prothrombin time and decrease of plasma fibrinogen level indicates moderate liver impairment. The therapeutic implication is an increased risk of bleeding and the need to give fresh frozen plasma to patients who are bleeding or require any interventional procedure.
BIOCHEMISTRY

- Serum electrolytes, blood urea nitrogen and serum creatinine
- Serum bilirubin, albumin, aminotransferases, alkaline phosphatase and γ-glutamyltranspeptidase. These indices assess the severity and activity of the underlying liver disease
SEROLOGY

• Hepatitis A, B and C serologies should be carried out routinely
• Antimitochondrial antibody (primary biliary cirrhosis);
• Antinuclear antibody (for evidence of autoimmune hepatitis);
• Ceruloplasmin for Wilson’s disease; and
• $\alpha_1$-antitrypsin to reveal deficiency.
ENDOSCOPY

• All patients confirmed or suspected of having cirrhosis should have a screening upper gastrointestinal endoscopy. If varices are identified at screening, patients should be started on appropriate prophylactic pharmacologic therapy to reduce the risk of an initial bleed.

• For a patient who has cirrhosis and has an upper gastrointestinal bleed, endoscopy is the first step in evaluation of the bleeding site and in making decisions on further management.

• The classification of the severity of varices is based on size, extent and the ‘color signs’.
RADIOLOGIC IMAGING

- The initial imaging study is ultrasound with Doppler evaluation of the major vessels. An ultrasound scan of the liver should inspect overall morphology; it is the initial screening procedure for focal intrahepatic lesions that may suggest hepatocellular carcinoma. The vessel imaging focuses on the main portal vein, the splenic vein, the hepatic veins and the infra- and intrahepatic inferior vena cava. It identifies most of the extrahepatic causes of portal hypertension.

- A CT scan or MRI of the abdomen may be indicated for some patients who have portal hypertension to evaluate the liver morphology in greater detail and to image the intra-abdominal venous system.
Fig. 27-8 Venous phase imaging of superior mesenteric artery injection demonstrating the superior mesenteric and portal veins.
LIVER BIOPSY

- Percutaneous biopsy
- Transjugular biopsy
- Laparoscopic biopsy
### Treatment Strategies for Esophagogastric Varices

<table>
<thead>
<tr>
<th>Strategy</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pharmacologic therapy</td>
</tr>
<tr>
<td>Endoscopic therapy</td>
</tr>
<tr>
<td>Tamponade</td>
</tr>
<tr>
<td>Decompression – radiologic, surgical</td>
</tr>
<tr>
<td>Devascularization operation</td>
</tr>
<tr>
<td>Liver transplantation</td>
</tr>
</tbody>
</table>

Fig.27-10  Treatment strategies for esophagogastric varices.
Prophylaxis to prevent the initial bleed

Treatment of acute variceal bleeding

Treatment to prevent recurrent variceal bleeding – first-line treatment,
   – second-line treatment

Treatment of end-stage liver disease

Fig. 27-11 Time points in the treatment of esophogastric varices.
PHARMACOLOGIC THERAPY

• The patient who has acute variceal bleeding may be treated initially with intravenous vasopressin and nitroglycerin or somatostatin or one of its analogs.

• Acute pharmacologic therapy is most easily administered using octreotide infused at 50mg/h. This agent is as effective as vasopressin and nitroglycerin given in combination, has fewer side effects and requires less monitoring.

• In the elective situation, noncardioselective β-blockers and the long-acting nitrates are the main drugs used to lower portal pressure. Portal pressure could be lowered by about 20% with propranolol (Oral pharmacologic therapy). A similar effect can be obtained with the long-acting nitrates.
ENDOSCOPIC THERAPY

• Endoscopic therapy has a definite role in acute variceal bleeding and is the first-line therapy in preventing rebleeding. The two main techniques are sclerotherapy or variceal banding. (1) Sclerotherapy involves the use of one of several agents to directly thrombose a varix or to create fibrosis in the mucosa overlying a varix; (2) In banding the varix is sucked into an applicator on the end of the flexible endoscope and a rubber band is fired to strangulate the varix; the bands slough off in 7–10 days with minimal scarring and less ulceration than with sclerotherapy.

• Endoscopic banding is more effective, resulting in earlier obliteration of varices and fewer complications than endoscopic sclerotherapy.
TAMPONADE

• This is rarely indicated in the 1990s, but may be needed for the 10% of patients whose acute bleeding is not controlled endoscopically. The Sengstaken–Blakemore tube has both gastric and esophageal balloons.

• The tube must first be fully introduced into the stomach. The gastric balloon should then be inflated to 300–350ml and pulled up into the gastric fundus. Occasionally, persistent bleeding may occur in the esophagus, requiring inflation of the esophageal balloon to 40mmHg.

• The tube should not be left in place for more than 12–24 hours and should only be used as a temporary measure while the patient is resuscitated and prepared for either repeat endoscopic therapy or decompression.
## Methods of Decompression for Portal Hypertension and Gastroesophageal Varices

<table>
<thead>
<tr>
<th>Description</th>
<th>Details</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>TIPS</strong></td>
<td></td>
</tr>
<tr>
<td>Nonoperative, radiologic</td>
<td></td>
</tr>
<tr>
<td>Portosystemic surgical shunts</td>
<td></td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td>Portacaval (using portal vein)</td>
</tr>
<tr>
<td></td>
<td>Mesocaval, mesorenal, mesoatrial (using superior mesenteric vein)</td>
</tr>
<tr>
<td><strong>Partial</strong></td>
<td>Portacaval or mesocaval (interposition graft of restricted diameter to allow some prograde portal flow)</td>
</tr>
<tr>
<td><strong>Selective variceal decompression</strong></td>
<td></td>
</tr>
<tr>
<td><strong>Distal splenorenal shunt</strong></td>
<td></td>
</tr>
</tbody>
</table>
Fig. 27-13 TIPS. The metallic shunt maintains the transparenchymal track, which has been made through the liver between the portal and hepatic veins.
Fig. 27-14  Side-to-side portacaval shunt with direct vein-to-vein anastomosis. The portal vein is dissected from the right side, posterior to the bile duct. The intrahepatic vena cava is fully mobilised. If there is a larger caudate lobe, an interposition graft may be used.
Fig.27-15 Partial portosystemic shunt with an 8mm graft between the portal vein and inferior vena cava. Exposure is the same as for the total portacaval shunt.
Fig. 27-16 Distal splenorenal shunt, with anastomosis of the splenic to the left renal vein. The gastric fundus, esophagus and spleen are decompressed. Portal hypertension and prograde portal flow are maintained.
DEVASCULARIZATION PROCEDURES

• These have the components of splenectomy, gastric and esophageal devascularization, and esophageal transection, with the goal of reducing inflow to bleeding varices. Their success depends upon the extent of the devascularization: the more extensive, the lower the subsequent rebleeding rate.

• Sufficient exposure: easy to perform splenectomy and to immobilize fully both the stomach and at least 7cm of the distal esophagus. pyloroplasty is necessary.

• The upper two-thirds of the lesser curve of the stomach should be devascularized. The whole of the greater curve should be devascularized from the pylorus to the gastroesophageal junction.
Fig. 24.17 Gastroesophageal devascularization interrupts all variceal inflow to most of the stomach and the distal 7cm of the esophagus. Splenectomy and desvascularization of the greater curve of the stomach complete the procedure.
This is part of the surgical repertoire of managing patients who have portal hypertension and variceal bleeding. The indication for transplant remains end-stage liver disease, and for Child’s class C patients this treatment has altered the long-term outcome.
MANAGEMENT OF VARICEAL BLEEDING AT DEFINED TIME POINTS

• **PROPHYLACTIC THERAPY**
  Pharmacologic therapy to prevent the first bleed has been most widely studied using b-blockers.

• **ACUTE VARICEAL BLEEDING**
  Balloon tamponade
  Decompression
PREVENTION OF RECURRENT VARICEAL BLEEDING

The principles for prevention of rebleeding can be summarized as:

• first-line treatment with pharmacologic and endoscopic therapy;
• second-line treatment with variceal decompression; and
• liver transplantation for end-stage liver disease.
Fig. 27-18  Management of variceal bleeding. DSRS, distal splenorenal shunt.
MANAGEMENT OF EXTRAHEPATIC PORTAL VEIN THROMBOSIS

- normal liver function;
- control bleeding: endoscopic therapy; decompression; devascularization; splenectomy
- identify the etiology: angiography is more important
ASCITES
PATHOGENESIS

Ascites results from a combination of:

• portal hypertension;
• altered renal sodium and water handling;
• hypoalbuminemia.
MANAGEMENT

• Ascites is managed primarily by medical therapy
• If ascites is refractory to medical management, the next line of therapy is paracentesis. Large-volume paracentesis
• The primary surgical treatment for intractable ascites is liver transplantation because the patient has ‘decompensated’ liver disease.
BUDD–CHIARI SYNDROME

<table>
<thead>
<tr>
<th>Cause</th>
<th>Proportion of cases (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Myeloproliferative disorder</td>
<td>50</td>
</tr>
<tr>
<td>Idiopathic</td>
<td>20</td>
</tr>
<tr>
<td>Tumor</td>
<td>10</td>
</tr>
<tr>
<td>Estrogen use, pregnancy</td>
<td>10</td>
</tr>
<tr>
<td>Hypercoagulable state</td>
<td>5</td>
</tr>
<tr>
<td>Paroxysmal nocturnal hemoglobinuria</td>
<td>5</td>
</tr>
</tbody>
</table>
PATHOPHYSIOLOGY

• The sequence of outflow obstruction, sinusoidal dilatation and congestion, hepatocyte necrosis from the increased pressure and progressive liver damage produce the syndrome.

CLINICAL PRESENTATION

• This is usually subacute with relatively mild symptoms and signs of ascites, hepatomegaly and right upper quadrant abdominal pain.
Fig. 27-22  Evaluation and management of Budd–Chiari syndrome. OLT, orthotopic liver transplantation.
Thank you!