Portal Hypertension

Portal hypertension refers to abnormally high pressure in the hepatic portal vein. Clinically significant portal hypertension is defined as an hepatic venous pressure gradient of 10 mm Hg or more.[1]

See related articles dealing with Ascites, Cirrhosis, Hepatic Encephalopathy, Hepatorenal Syndrome, liver failure and oesophageal varices, listed below under 'Complications'.

Aetiology

Causes can be divided into prehepatic, hepatic and posthepatic. Cirrhosis is the most common cause.

Causes of portal hypertension

Prehepatic - blockage of the portal vein before the liver

- Congenital atresia or stenosis.
- Portal vein thrombosis (idiopathic, umbilical and portal sepsis, malignancy, hypercoagulable states, pancreatitis).
- Splenic vein thrombosis.
- Extrinsic vein thrombosis.
- Extrinsic compression - eg, tumours.

Hepatic

- Cirrhosis.
- Chronic hepatitis.
- Schistosomiasis.
- Myeloproliferative diseases.
- Idiopathic portal hypertension.
- Granulomata - eg, sarcoid.
- Nodular (nodular regenerative hyperplasia, partial nodular transformation).
- Toxins (arsenic, vinyl chloride).
- Fibropolycystic disease (including congenital hepatic fibrosis).

Posthepatic - blockage of hepatic veins or venules

- Budd-Chiari syndrome (hepatic vein obstruction).
- Constrictive pericarditis.
- Right heart failure.
- Veno-occlusive disease of the smaller hepatic veins/venules (due to ingestion of pyrrolizidine alkaloids; antileukaemic drugs, radiation).
- Sclerosing hyaline necrosis.

Other causes

- Increased hepatic blood flow:
  - Increased splenic blood flow - eg, massive splenomegaly.
  - Hepatoportal arteriovenous fistula.
- Idiopathic (a diagnosis of exclusion).

Left-sided (sinistral) portal hypertension
- Rare. It is confined to the left side of the portal system.
- It may present as bleeding from gastric varices.
- Usually it is due to pathology involving the splenic vein or the pancreas.

**Pathophysiology**

Portal hypertension develops due to:

- Increased vascular resistance in the portal venous system - from various mechanical causes (above), and also as an active process in which liver damage activates stellate cells and myofibroblasts, contributing to the abnormal blood flow patterns.
- Increased blood flow in the portal veins - from splanchnic arteriolar vasodilatation, caused by an excessive release of endogenous vasodilators.

The raised portal pressure opens up venous collaterals, connecting the portal and systemic venous systems. These occur in various sites:

- Gastro-oesophageal junction - producing varices which are superficial and easily bleed.
- Anterior abdominal wall:
  - Via the umbilical vein - visible as caput medusae radiating from the umbilicus.
  - May also occur where adhesions exist between abdominal viscera and the parietal peritoneum, or at sites of stomas or previous surgery.

- Anorectal junction - rarely cause bleeding.
- Veins from retroperitoneal viscera - communicate with systemic veins on the posterior abdominal wall.

Other patterns of blood flow:

- If individual tributaries of the portal vein are thrombosed, this causes local venous hypertension. With splenic vein block, oesophageal and gastric varices may result.
- In Budd-Chiari syndrome (hepatic vein occlusion), collaterals open up within the liver; blood tends to be diverted through the caudate lobe whose short hepatic veins drain directly into the inferior vena cava.

Portosystemic venous anastomoses can cause encephalopathy, possibly due to various 'toxins' bypassing the liver's 'detoxification' process.

Circulatory disturbances:

- Portal hypertension and cirrhosis produce a hyperdynamic circulation, with splanchnic vasodilatation, increased cardiac output, arterial hypotension, and hypervolaemia.
- There is salt and water retention, ascites and hyponatraemia.

**Presentation**

**History**

For causes of liver disease:

- History of jaundice.
- Alcohol consumption.
- Blood transfusion, especially abroad; lifestyles that predispose to hepatitis B or hepatitis C.
- Family history - eg, Wilson's disease or hereditary haemochromatosis.

For complications of portal hypertension:

- Haematemesis or melaena - suggest bleeding varices.
- Lethargy, irritability and changes in sleep pattern - suggest encephalopathy.
- Increased abdominal girth, weight gain - suggest ascites.
Abdominal pain and fever - suggest spontaneous bacterial peritonitis. Pulmonary involvement is common in patients with portal hypertension.\[2\]

Examination

Signs of portal hypertension:

- Dilated veins in the anterior abdominal wall and caput medusae (tortuous collaterals around the umbilicus). A venous hum, loudest during inspiration, is sometimes heard over large upper abdominal collaterals.
- Splenomegaly.
- Ascites.

Signs of liver failure:

- Jaundice, spider naevi, palmar erythema.
- Confusion, liver flap and fetor hepaticus are signs of encephalopathy.
- Signs of hyperdynamic circulation: bounding pulse, low blood pressure, warm peripheries.
- Enlarged or small liver.
- Gynaecomastia and testicular atrophy.

Investigations

- Blood tests:
  - LFTs, U&Es, glucose, FBC, clotting screen.
  - Investigations for liver disease if the cause is not known - eg, ferritin (for haemochromatosis), hepatitis serology, autoantibodies, alpha-1 antitrypsin (for alpha-1-antitrypsin deficiency), ceruloplasmin (for Wilson's disease).

- Scans:
  - Abdominal ultrasound - for liver and spleen size, ascites, portal blood flow and thrombosis of the portal or splenic veins.
  - Doppler ultrasound - can show direction of flow in blood vessels.
  - CT scan, especially spiral CT, may show portal vasculature - can be useful if ultrasound was inconclusive.
  - MRI scan - gives similar information to CT.
  - Elasticity measurement (FibroScan®) - a new technique based on the velocity of an elastic wave via an intercostally placed transmitter. Results correlate with liver stiffness and so with fibrosis.\[3\]

- Endoscopy - for oesophageal varices - essential for those with suspected portal hypertension. Varices indicate portal hypertension, but their absence does not exclude it.
- Portal hypertension measurement:\[4\]
  - Portal pressure is indirectly measured in clinical practice by the hepatic venous pressure gradient (HVPG).
  - Normal HVPG values are <5 mm Hg. HVPG >10 mm Hg predicts the development of oesophageal varices.
  - However, HVPG is moderately invasive and its clinical role is uncertain.

- Liver biopsy - if indicated, may help diagnose the underlying cause.

Vascular imaging:

- The site of the portal venous block can be demonstrated by examining the venous phase of a coeliac or superior mesenteric arteriogram, by splenic portography following injection of dye into the splenic pulp, or by retrograde portography via a hepatic vein.
- Hepatic venography is helpful when hepatic vein block or idiopathic portal hypertension is suspected.

Management

The portal hypertension itself is difficult to treat effectively, except by treating the underlying cause (where possible) and liver transplantation (if indicated and feasible).
Portal venous pressure can be reduced by beta-blockers ± nitrates and by using shunt procedures to create an anastomosis between the portal and hepatic veins (below).

- Conservative management includes salt restriction and diuretics.\[^5\]
- Some pulmonary vasoactive drugs (eg, epoprostenol) have become routine in the treatment of patients with idiopathic pulmonary hypertension.\[^6\]
- Bleeding from oesophageal varices is a life-threatening complication of portal hypertension. Primary prevention of bleeding in patients at risk for a first bleeding episode is therefore a major goal. Medical prophylaxis consists of non-selective beta-blockers (eg, propranolol or carvedilol).\[^7\]
- Continuous infusion of terlipressin reduces portal venous pressure stably and may become an alternative to traditional bolus injection.\[^8\]
- Transjugular intrahepatic portosystemic shunt (TIPS) has become a mainstay treatment option for the management of portal hypertension-related complications in liver cirrhosis.\[^9\]
- Because of high perioperative mortality, transplantation should be avoided in those patients with portopulmonary hypertension who have severe pulmonary hypertension that is refractory to medical therapy.\[^10\]

The following gives an overview of treatments used on the portal vascular system:

**Drug treatments**\[^4\]

**Beta-blockers**
- Non-selective beta-blockers reduce portal pressure in many patients:
  - They reduce rates of bleeding and re-bleeding in patients with oesophageal varices.
  - They may also protect against spontaneous bacterial peritonitis (perhaps through increasing intestinal transit).

- Carvedilol (a non-selective beta-blocker with anti-alpha1-adrenergic effects) showed promising results in a study, and may have a role in preventing variceal bleeds.

**Nitrates**
- Added to beta-blocker therapy, they contribute to reducing portal pressure and may reduce rates of variceal re-bleeding.

**Vasoactive drugs**
- Terlipressin and octreotide are used to assist the control of acute variceal bleeding.\[^11\]

**Endoscopic procedures**\[^4\]
- Endoscopy - to detect and monitor oesophageal varices.
- Endoscopic vein ligation - to prevent bleeding of oesophageal varices.
- For gastric varices with acute bleeding, endoscopic variceal obturation with tissue adhesives (eg, cyanoacrylate) is effective - but there are recognised complications (mucosal ulceration and thromboembolism).

**Transjugular intrahepatic portosystemic shunt (TIPS)**\[^12, 13\]
This is a radiological procedure, connecting the portal and hepatic veins using a stent. The purpose of a TIPS is to decompress the portal venous system, to prevent re-bleeding from varices or to reduce the formation of ascites.

However, there are potential complications - hepatic encephalopathy and deteriorating liver function. The stent may stenose; it requires follow-up and may require repeat procedures. There are various contra-indications, detailed in recent guidelines.\[^4\]

TIPS is an established treatment option for:
Ascites - for patients requiring repeated and frequent paracentesis.
Oesophageal variceal bleeding refractory to medical treatment (acute bleeding or secondary prevention).
Bleeding from non-oesophageal varices - e.g., gastric varices.

TIPS may also have a role in treating:

- Hepatorenal syndrome.
- Hepatic hydrothorax.
- Hepatopulmonary syndrome.
- Budd-Chiari syndrome.

Surgical procedures

Surgical portosystemic shunts: \(^{[14]}\)

- These require major surgery and an experienced surgeon. They are less likely to stenose than TIPS and can be used where TIPS is not feasible.
- Shunts may be total, partial or selective.

Devascularisation procedures:

- Include gastro-oesophageal devascularisation, splenectomy and oesophageal transection.
- Generally used where other therapies are unsuitable.

Management of rectal varices

- These are common in patients with portal hypertension but don't usually bleed. They are located at the anorectal junction.
- If they bleed, suggested treatment is similar to that used for upper gastrointestinal varices - using drugs to reduce portal pressure, endoscopic banding and TIPS if bleeding persists. \(^{[15]}\)

Complications \(^{[3]}\)

The complications of portal hypertension ± cirrhosis include:

- Bleeding from gastric or oesophageal varices - the most common complication of portal hypertension. Gastric varices occur in around 20% of patients with portal hypertension, mostly secondary to liver cirrhosis. \(^{[16]}\)
- Ascites and its complications:
  - Spontaneous bacterial peritonitis.
  - Hepatorenal syndrome (a complication of cirrhosis with ascites).
  - Hepatic hydrothorax. \(^{[17]}\)
- Pulmonary complications: \(^{[17]}\)
  - Portopulmonary hypertension (pulmonary arterial hypertension complicating portal hypertension in patients with liver disease).
  - Hepatopulmonary syndrome (a triad of hepatic dysfunction, hypoxaemia and extreme vasodilation in the form of intrapulmonary vascular dilations).
- Liver failure.
- Hepatic encephalopathy.
- Cirrhotic cardiomyopathy. \(^{[18]}\)

For specific management, see separate articles Ascites, Cirrhosis, Hepatic Encephalopathy, Hepatorenal Syndrome, Liver Failure and Oesophageal Varices.

Prognosis
This depends on the prognosis of the underlying disease, and on the outcome of any complications such as variceal bleeding.

The Child-Pugh classification system indicates prognosis in some types of liver disease such as cirrhosis.[19]

<table>
<thead>
<tr>
<th>Criterion</th>
<th>Score 1 point</th>
<th>Score 2 points</th>
<th>Score 3 points</th>
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<tbody>
<tr>
<td>Serum albumin (g/L)</td>
<td>&gt;35</td>
<td>28-35</td>
<td>&lt;28</td>
</tr>
<tr>
<td>Serum bilirubin (total) [3]</td>
<td>&lt;34 μmol/L (&lt;2 mg/dL)</td>
<td>34-50 μmol/L (2-3 mg/dL)</td>
<td>&gt;50 μmol/L (&gt;3 mg/dL)</td>
</tr>
<tr>
<td>International Normalized Ratio (INR)</td>
<td>&lt;1.7</td>
<td>1.7-2.2</td>
<td>&gt;2.2</td>
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<tr>
<td>Ascites</td>
<td>Absent</td>
<td>Controlled medically</td>
<td>Poorly controlled</td>
</tr>
<tr>
<td>Encephalopathy</td>
<td>Absent</td>
<td>Controlled medically</td>
<td>Poorly controlled</td>
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</tbody>
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A score of 5-6 is class A (life expectancy 15-20 years); a score of 7-9 is class B (life expectancy 4-14 years); a score of 10-15 is class C (life expectancy 1-3 years). This aligns with a perioperative mortality (for abdominal surgery) of 10%, 30%, and 80% respectively.

Survival rates according to Child-Pugh class are: one-year survival in class A is 100%; class B 81%; class C 45%.

Other scoring systems have been developed, including:

- Model for End-stage Liver Disease (MELD). [3]
- Additions to MELD such as:
  - MELD-Na (incorporates serum sodium). [20]
  - MELD-ICG (incorporates indocyanine green clearance). [20]

Variceal haemorrhage, especially from oesophageal varices, is the most common complication associated with portal hypertension. Almost 90% of patients with cirrhosis develop varices, but only 30% of varices bleed. Variceal haemorrhage has a one-year mortality rate of 40%. [21]

Further reading & references


11. Management of acute upper and lower gastrointestinal bleeding; Scottish Intercollegiate Guidelines Network - SIGN (September 2008)

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