INTRODUCTION

The portal vein is formed by the confluence of the splenic vein with the superior mesenteric vein and its formation mostly occurs behind the pancreas in the retro peritoneum. It transports the blood mainly from the gastro-intestinal tract and the spleen to the liver. Seventy percent of the total blood supply to the liver is contributed by the portal vein while the hepatic artery contributes to the remaining thirty percent. The portal venous system is the only venous system in our body, which begins with capillaries and ends with capillaries. The intrahepatic branches of the portal vein terminate in small vessels that supply the hepatic sinusoids. Embryologically, the systemic veins of our body develop from the intra-embryonic anterior and posterior cardinal veins while the portal system develops from the extra-embryonic vitelline and umbilical veins, which drain from the yolk sac and the placenta.

DEFINITION

Portal hypertension could be defined as an increase in the intravascular pressure within the portal vein of over 11 mm of mercury as measured directly or a splenic pulp pressure of over 16 mm of mercury.

PATHOPHYSIOLOGY

A rise in the portal pressure leads to splenomegaly and the development of natural porto-systemic shunts at the following sites:

- Lower end of the oesophagus and cardia through the gastro-oesophageal veins
- The anal canal via the hemorrhoidal veins
- In the falciform ligament via the umbilical veins
- In the abdominal wall and retroperitoneum

CLASSIFICATION
The etiology of portal hypertension in children is classified as:

- **Cirrhotic** - e.g. biliary atresia, cystic fibrosis
- **Non-cirrhotic**
  - **Pre-hepatic** - e.g. portal vein thrombosis, splenic vein thrombosis
  - **Intra-hepatic**
    - Presinusoidal – e.g. noncirrhotic portal fibrosis, congenital hepatic fibrosis, sclerosing cholangitis, schistosomiasis
    - Parasinusoidal – e.g. fatty liver, nodular hyperplasia, alcoholic hepatitis
    - Postsinusoidal – e.g. veno-occlusive disease of liver, hepatic vein thrombosis
  - **Supra-hepatic** – IVC web (Budd-Chiari syndrome), constrictive pericarditis

**Non cirrhotic Portal Fibrosis** (NCPF) and extrahepatic portal vein thrombosis are two diseases that are common in developing countries and most often present with features of portal hypertension and not of parenchyma dysfunction. The patients with NCPF are generally young and come from low socio-economic status.

**EHPVO** is the commonest cause of portal hypertension in children in India (80%-90% of cases of portal hypertension) is a vascular disorder of the liver and is defined as obstruction of the extrahepatic portal vein with or without involvement of the intrahepatic portal veins or splenic or superior mesenteric veins, isolated occlusion of splenic vein or superior mesenteric vein does not constitute EHPVO. The liver function tests in such patients are essentially normal. A confirmation of portal vein occlusion may be obtained by ultrasound demonstration of collateral venous channels at the porta hepatitis replacing the portal vein. Approximately 40% of these patients have a history of umbilical vein catheterization or abdominal sepsis in the neonatal period but the venous occlusion in the majority appears to be congenital in origin.

The most frequent cause of cirrhosis in childhood is **biliary atresia**. Besides this metabolic liver diseases are other common medical conditions leading to cirrhosis of the liver. Many of these patients have stigmata of underlying disease and the diagnosis of portal hypertension is not difficult.

**Congenital hepatic fibrosis** may also present with an acute hematemesis and normal liver function tests but the clinical features include hepatomegaly. A liver biopsy shows bands of fibrous tissue joining the portal tracts and this condition may be associated with polycystic disease and other renal disorders. There are several reports of portal hypertension in children presenting with hematemeses and splenomegaly in whom the liver histology is normal and in whom the portal vein is patent.

**MANAGEMENT**

Management of Portal Hypertension includes management of variceal hemorrhage, hypersplenism and other complications such as portal biliopathy, ascites, malabsorption etc.
The survival of the children with portal hypertension depends almost entirely on the etiology. Recent reports show that oesophageal varices in childhood are well controlled with either injection sclerotherapy or porto-systemic shunting and both methods have their advocates. Patients with portal vein obstruction and normal liver histology can be expected to live normal lives providing the oesophageal varices are under control.

A) Management of Variceal bleed

Acute variceal bleeding, particularly in young infants, can pose problems in management and delay in immediate management could prove fatal for a child.

Medical measures include

- **Airway protection**
- **Hemodynamic resuscitation**
- **Pharmacologic therapy**
  - **Vasopressin** - The intravenous infusion of (0.2 – 0.4 units/1.73 m / min) which may arrest the bleeding. Vasopressin or its precursor, glypressin may be used alone or in combination with nitrates to reduce the portal venous pressure. Unfortunately, these agents have side-effects related to systemic vaso-constriction like headache, nausea and abdominal cramps.
  - **Somatostatin** - It reduces splanchnic blood flow and portal pressure with minimal side-effects, but it has a short half life of less than 3 minutes. Octreotide, a long acting analog of somatostatin, has a plasma half-life of more than 1 hour. Although the effectiveness of octreotide has been studied in a small number of children, its safety and side-effect profile have encouraged its use in cases of acute variceal bleeding.
- **Endotherapy**
  - **Injection sclerotherapy** - However continued bleeding may be controlled with injection sclerotherapy but the small size of the pediatric endoscope channels can limit the clearance of blood from within the oesophagus. In addition to the above difficulties, there is an added risk of needing general anesthesia in a child with a compromised consciousness.
  - **Variceal banding** - This technique, which involves application of an elastic band to a variceal column, is done through flexible upper gastro-intestinal endoscopes. The strangulated varix, subsequently, thromboses and sloughs. Usually 3-6 bands are applied at each session. Multi-band devices allow the application of several bands without the need for reloading. Treatment is performed initially at 1 to 2 weekly intervals, extending to monthly intervals once the larger varices are treated. The incidence of oesophageal stricture and systemic side-effects is lower with this treatment modality. At present, equipment limitations make this technique difficult to use in small children less than 2 years of age.
  - **Glue Injection** : Endoscopic variceal occlusion with tissue adhesives such an n-butyl-cyanoacrylate or thrombin is more effective for acute fundal gastric varices in adults.
- **Sangstaken-Blackmore (S-B tube) compression balloon** - It may be life saving when there is a failure of visualization of the varices due to overwhelming hemorrhage. However, the dangers of this instrument cannot be overemphasized. Correct placement of the gastric balloon must be checked with X-ray control in order to avoid the inflation within the lumen of the oesophagus. This accidental inflation with the oesophagus may result in oesophageal rupture or suffocation.
from airway obstruction. Inflation of the gastric balloon and moderate prolonged traction achieved by securing the S-B tube to the side of the face with an adhesive tape is usually sufficient to stop the bleeding. It is rarely necessary to inflate the oesophageal balloon present on the standard instrument. Balloon deflation is performed 18 to 24 hours later and this is followed immediately with endoscopic variceal injection.

**Long term management of esophageal varices**

- Endotherapy
- Surgery – Portosystemic shunts, liver transplantation

**Injection sclerotherapy for long-term treatment.**

Injection sclerotherapy was suggested for the treatment of oesophageal varices in children because of failures and complications of primary surgery. Portosystemic shunt thrombosis and rebleeding, the hazards of splenectomy in children and long term risks of encephalopathy all encouraged an alternative therapy. Controlled trials in patients confirmed that early endoscopic sclerotherapy after the onset of bleeding significantly reduced the risk of rebleeding and may prolong survival in the cirrhotic. Injections are performed through a flexible upper GI endoscope under general anesthesia /deep sedation. Intravenous sedation has been used occasionally in older children. A variety of sclerosants are available including ethanolamine oleate, sodium tetradecyl sulphate, sodium morrhuate, phenol in almond oil and polidocanol. The injections are given either intra or para-variceal and are mostly given into the cardia and lower 3 cm of the oesophagus. A maximum of 3 ml is injected into each varix to a maximum of 5 to 20 ml per session depending on the age and the size of the patient. A naso-gastric tube is inserted in small infants to control the degree of gastric distension. The initial 3 injections are given at weekly intervals and subsequent treatments on a monthly basis until the varices are obliterated.

Mild symptoms of retrosternal discomfort and transient fever are common after endoscopic sclerotherapy. The variceal haemorrhage may recur, particularly between the first 2 or 3 treatments and oesophageal ulceration may be followed by stricture formation and dysphagia. Rare serious complications have included broncho-oesophageal fistula, chylothorax and pericarditis. One case of paraplegia has been reported from injection of segmental spinal vessels.

An analysis of seven reports published since 1984 of the results of sclerotherapy in 248 children shows a mortality rate of 3 percent and a rebleed rate of 12 percent. The rebleed rate in a series of 7 reports of surgery for portal hypertension (1980 – 86) was 14 percent.

**Transjugular intrahepatic port-systemic shunt (TIPS)**

The indications of TIPS in children include uncontrolled variceal bleeding especially the ones who are awaiting liver transplantation. Some patients of Budd-Chiari syndrome or intractable ascitis may also benefit by this procedure. Portal vein thrombosis, bacterial sepsis and coagulopathy are contraindications to TIPS.

This intervention involves insertion of an expandable metallic stent from the hepatic to the portal vein through the percutaneous tranjugular route under radiological guidance. Under fluoroscopic control, a guidewire is passed into a hepatic vein. A needle is then advanced over a guidewire into the hepatic vein
and thence to the portal vein. A balloon catheter is subsequently used to dilate the intrahepatic tract and the stent is deployed.

**B) Management of Hypersplenism**
Although a reduction in one or more hemopoetic cell lines is common in EHPVO, symptomatic hypersplenism is found in less than 5% of the patients. Splenomegaly along with shunt surgery may be indicated.

**C) Management of Portal Hypertensive Gastropathy**
Treatment is directed at decreasing portal pressure. Propranolol had been studied in adults. Endoscopic thermal coagulation is not effective for diffuse bleeding. Liver transplantation is indicated for decompensated liver disease.

**D) Management of Portal Biliopathy**
Portal biliopathy refers to abnormalities of the extrahepatic and intrahepatic bile ducts with or without anomalies of gall bladder wall in patients who have portal hypertension. The changes include indentations of paracholedochal collaterals on the bile duct, localized strictures, angulation of ducts, displacement of ducts and stones in the common bile duct and focal narrowing, dilatations, irregular walls, and clustering of intrahepatic branches in the hepatic ducts. Although biliopathy is seen 80% to 100% of cases, only a few patients are symptomatic. Symptomatic patients usually are adults, which indicates that portal biliopathy is a progressive disease. Complications, such as cholangitis, secondary biliary cirrhosis, gall stones, hemobilia, hypoalbuminemia, and coagulation disturbances, have been reported.

Symptomatic portal biliopathy could be managed by endotherapy (stone extraction, stricture dilatation, stenting) or surgery (shunt surgery)