

PORTAL HYPERTENSION IN CHILDREN

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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 retroperitoneum. 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. 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 haemorrhoidal veins
- In the falciform ligament via the umbilical veins
- In the abdominal wall and retroperitoneum

The diagnosis of portal hypertension should be suspected in a child after the occurrence of any large gastro-intestinal bleed. In this age group, oesophageal varices are the most likely cause for such an event. Variceal bleeding is associated with a mortality rate of 5 – 9 percent in children with portal vein obstruction but there is a higher risk of death of those with cirrhosis.

Classification:

The aetiology of portal hypertension in children is classified as: -

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| • | Cirrhotic - e.g. biliary atresia, cystic fibrosis |
| • | Non- cirrhotic |
| | • Pre-hepatic - e.g. portal vein thrombosis |
| | • Intra-hepatic – |
| | • Presinusoidal – e.g. congenital hepatic fibrosis |
| | • Parasinusoidal – e.g. fatty liver, nodular hyperplasia |

- Postsinusoidal – e.g. veno-occlusive disease of liver

- Supra-hepatic – e.g. Budd-Chiari syndrome

The most frequent surgical cause of cirrhosis in childhood is biliary atresia . Besides this cause alpha-1- antitrypsin deficiency and metabolic liver diseases are the commonest medical conditions leading up to cirrhosis of the liver. Many of these patients have the stigmata of their underlying disease and the diagnosis of portal hypertension is not difficult.

The diagnosis may not be so obvious in children with non-cirrhotic portal hypertension. Portal vein thrombosis (extra-hepatic portal hypertension), for example, may present within the first five years of life as a major haematemesis with only splenomegaly and a reduced platelet count as clues to the diagnosis. Many affected patients have no documented cause for their portal hypertension. However, at times, a history of umbilical vein canulation, abdominal infection, trauma or pancreatitis may be responsible for the portal thrombosis. 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 in the porta hepatis 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.

Congenital hepatic fibrosis may also present with an acute haematemesis 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 haematemesis and splenomegaly in whom the liver histology is normal and in whom the portal vein is patent.

Further studies have shown a subendothelial thickening of intrahepatic branches of the portal vein causing presinusoidal obstruction to portal blood flow within the liver leading to the formation of collateral venous channels in the porta hepatis. The condition has been known under a variety of names such as non-cirrhotic portal hypertension but the best is perhaps ' hepato-portal sclerosis'. The aetiology is unknown.

Suprahepatic obstruction caused by either a web in the inferior vena cava above the entrance of the hepatic veins or thrombotic occlusion of the hepatic veins (Budd-Chiari syndrome) is extremely rare in childhood. The clinical features may be mimicked by constrictive pericarditis but echocardiography and venography should make the diagnosis clear.

Clinical features :

In chronic liver disease, the presentation of portal hypertension is mainly as an abdominal mass due to splenomegaly. This is usually noted in slightly older children (around 8 years of age). Encephalopathy and abdominal distension due to ascitis may also complicate the haematemesis in patients with cirrhosis. Growth retardation is a well-recognized complication of cirrhosis and portal hypertension is a contributory factor to it due to mucosal oedema and lymphatic congestion leading to malabsorption and protein losing enteropathy.

However, in portal venous occlusion, the presentation is usually in younger children (5 years) with acute episode of upper or lower gastro-intestinal bleeding. This may or may not be accompanied by splenic enlargement depending on the blood loss due to the haematemesis or malena. Anorectal varices and haemorrhoids are identified in almost two-thirds of patients with portal venous occlusion.

In patients with Budd-Chiari syndrome, intractable ascitis with hepatomegaly is the usual initial presentation. Jaundice is variable.

Investigations:

Blood investigations

Hypersplenism may result in anaemia and reduced WBC and platelet count. Plasma concentrations of procoagulant and anticoagulant proteins may be reduced in portal venous thrombosis or Budd-Chiari syndrome. Biochemical liver function tests are abnormal in cirrhosis, but are rarely deranged in portal venous occlusion. Serum albumin levels are depleted in acute variceal bleed.

Ultrasound scan of abdomen

Large collateral veins, portal cavernoma and splenomegaly, which are the features of portal hypertension, may be identified on sonography of abdomen. Doppler studies provide the information about the direction, velocity and waveform characteristics of portal blood flow. In cirrhosis of liver, the maximum velocity of the blood flow in the main portal trunk is inversely correlated with the severity of the liver disease. In Budd-Chiari syndrome, Doppler sonography of the hepatic veins and the inferior vena cava can be conclusive. Ascitis which is a notable feature of Budd-Chiari syndrome and of cirrhosis of liver can be definitely identified and quantified on ultrasonography.

Upper and lower gastro-intestinal endoscopy

This modality is useful in the diagnosis and treatment of varices of the oesophagus, stomach, the proximal duodenum and the ano-rectal area. However, it may be safer and easier if the procedure is performed under a general anaesthesia. Various grading systems are used in the assessment of oesophageal varices. Smaller varices are bluish with a relatively thick mucosal covering while the larger ones may have signs of recent or impending bleed like 'cherry red spots' or 'varices on varices'. Portal congestive gastropathy is characterized by mucosal hyperaemia with dilated submucosal veins.

CT angiography and MRI scan

The above modalities are increasingly used in the diagnosis of Budd-Chiari syndrome and to identify liver lesions associated with portal hypertension like focal nodular regenerative hyperplasia. MR angiography is recently used as a non-invasive alternative to conventional angiography to delineate porto-mesenteric venous anatomy.

Angiography

Inferior vena cavography with pressure measurements is valuable in patients with Budd-Chiari syndrome in whom hepatic venography can be used to assess hepatic venous patency. Balloon dilatation can be undertaken of inferior vena caval membrane or short segment narrowing of the hepatic veins, which can prove therapeutic.

Treatment:

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) Treatment of the acute bleed

Acute variceal bleeding, particularly in young infants, can pose problems in management. A delay in immediate management could prove fatal for a child. Medical measures include blood transfusion and the intravenous infusion of vasopressin (0.2 – 0.4 units/1.73 m² / 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 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.

Continued bleeding may be controlled with injection sclerotherapy but the small size of the paediatric 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. The Sangstaken-Blackmore (S-B tube) compression balloon may be life saving when there is a failure of visualization of the varices due to overwhelming haemorrhage. 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.

B) 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 adult 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 anaesthesia with an endotracheal tube in place. Intravenous sedation has been used occasionally in older children. A variety of sclerosants are available including ethanalamine 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 a 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.

C) 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 upto three 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.

D) 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 transjugular 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.

E) Surgery – porto-systemic shunts

Endoscopic sclerotherapy with banding is an effective primary treatment modality of bleeding oesophageal varices in majority of children with reasonable liver function. However, surgical intervention is indicated for the following cases:

- Uncontrolled bleeding from the oesophageal varices not responding to at least 2 sessions of banding or sclerotherapy
- Bleeding gastric or ectopic varices not responding to endoscopic treatment
- Hypersplenism or massive symptomatic splenomegaly
- Lack of access to endoscopic treatment
- Symptomatic biliary obstruction due to choledochal varices
- Selected patients with Budd-Chiari syndrome

The great variety of surgical procedures advocated for the management of portal hypertension was reflected in a French multi-centre study of children treated both in European countries and North Africa . Thirty different operations, which included a range of porto-systemic shunts and various devascularization techniques, were reported from the treatment of 109 children. The variety of surgical techniques can be explained by both the varied pathology of the portal venous system and the preferences and experience of individual surgeons. Occlusion of the portal system in the extrahepatic portal hypertension, for example, may affect the portal vein alone or may involve either the splenic or the superior mesenteric veins. At least 20 percent of these patients are not suitable for any form of shunt surgery and may undergo some type of devascularization with or without oesophageal transection. Unfortunately, devascularisation procedures do have a high incidence of rebleeding of up to 23 percent.

The construction of a shunt between the superior mesenteric vein and the inferior vena cava using a segment of the internal jugular vein seems to offer the best combination of long term patency and the lowest incidence of rebleeding. The rebleed rate was only 6 percent in four series of cases published since 1981.

The recent introduction of the mesenterico-left portal (Rex) shunt is likely to broaden the indications once more for shunt surgery as the primary treatment for children with portal venous occlusion. This shunt utilizes an interposition graft between the superior mesenteric vein and the intra-hepatic portion of the left portal vein, which is identified in the Rex recessus adjacent to the falciform ligament. By restoring hepatic portal blood flow and correcting portal hypertension, this technique is more physiological and obviates the potential disadvantages of the porto-systemic shunts.

The complications of porto-systemic shunting are not only concerned with rebleeding. Deterioration of liver function and hepatic encephalopathy are further hazards, particularly in children with cirrhosis. In a recent series of 37 children, 1 died in the early postoperative

period with encephalopathy and 9 out of 31 with a patent shunt showed deterioration of mental function during a mean follow-up period of 5 years.

Porto-systemic shunt surgery should be regarded as a complementary therapy to endoscopic treatment. Porto-mesenteric venous anatomy does not permit successful shunt surgery in every child, and shunt thrombosis has been recorded with all types of shunts especially in smaller children.

Liver transplantation is the treatment of choice for children with variceal bleeding complicating end-stage chronic liver disease.

Conclusion:

The management of children with bleeding oesophageal varices is an extremely challenging task and demands a variety of complementary techniques, each of which may be limited by its applicability, efficacy and its complications. Endoscopic sclerotherapy and banding is highly effective and appears to be the treatment of choice for the initial management of oesophageal varices in children.

However, shunt surgery should be reserved for

- treatment of gastric or ectopic varices not accessible to sclerotherapy
- uncontrollable bleeding secondary to a complication of sclerotherapy
- treatment of severe hypersplenism or symptomatic splenomegaly
- treatment of children living in communities far away from adequate medical care and blood transfusion facilities.

Liver transplantation is the procedure of choice for patients with complications of portal hypertension associated with end-stage liver disease. The role of newer modalities like the TIPS and Rex shunt has yet to be conclusively proven.

References:

1. Howard ER, Stringer MD, Colombani PM. Surgery of the liver, bile ducts and pancreas in children. Second Edition Arnold Publishers, London 2002 pp 285 – 340
2. Bismuth H, Franco D, Alagille D. Portal diversion for portal hypertension in children. *Annals of Surgery* 1980; 192: 18 – 24
3. Fonkalsrud EW. Surgical management of portal hypertension in childhood. *Archives of Surgery* 1980; 115: 1042 – 1045
4. Howard ER, Stringer MD, Mowat AP. Assessment of injection sclerotherapy in the management of 152 children with oesophageal varices. *British Journal of Surgery* 1988; 75: 404 – 408
5. Stringer MD, Howard ER, Mowat AP. Endoscopic sclerotherapy in the management of oesophageal varices in 61 children with biliary atresia. *Journal of Pediatric Surgery* 1989; 24: 438 – 442
6. Fonkalsrud EW, Myers NA, Robinson MJ. Management of Extrahepatic Portal Hypertension in Children *Ann Surg.* 1974; 180(4): 487–491.
7. Poddar U, Thapa BR, Puri P et al Non-cirrhotic portal fibrosis in children *Indian Journal of Gastroenterology* 2000; 19(1): 12-13
8. De Ville de Goyet J, Alberti D, Clapuyt P et al. Direct bypassing of extrahepatic portal venous obstruction in children: a new technique for combined hepatic portal revascularization and treatment of extrahepatic portal hypertension. *Journal of Paediatric Surgery* 1998; 33:597-601
9. Heyman MB , LaBerge JM. Role of transjugular intrahepatic portosystemic shunt in the treatment of portal hypertension in paediatric patients. *Journal of Pediatric Gastroenterology and Nutrition* 1999; 29: 240 – 249
10. Price MR, Sartorelli KH, Karrer FM et al. Management of esophageal varices in children by endoscopic variceal ligation. *Journal of Paediatric Surgery* 1996; 31:

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