Surgical Aspects of Portal Hypertension

Sushmita N Bhatnagar (Pediatric Surgeon, Co-Incharge, Pediatric Liver Clinic)
P Shenoy (Pediatric Surgeon, Lecturer, Pediatric Liver Clinic)
Pediatric Liver Clinic, B.J.Wadia Hospital for Children.

INTRODUCTION

Surgical intervention for portal hypertension was the only treatment available in the earlier times and the mortality rates were very high with or without surgery. In today’s times, about 10-15% of patients require surgery.

The treatment of a child with portal hypertension is predicted first on the underlying pathophysiology of the disease and then on the severity of the symptoms. The prognosis of a child with liver disease and portal hypertension depends entirely on the hepatic reserve (1). With the advent and proven success of endoscopic treatment i.e. sclerotherapy, surgery no longer remains the first line of treatment in Portal hypertension. Extrahepatic portal hypertension is one of the most common entities wherein shunt surgery is performed with success.

Shunts have been attempted since 1877 wherein Eck ligated the portal vein and connected the visceral end to the vena cava. It commonly became known as Eck Fistula (2). In the second phase of developments in the surgical management which occurred in mid-1940’s a portacaval shunt was devised by Whipple and Blakemore.

INDICATIONS FOR SURGICAL INTERVENTION IN PHT

1. Uncontrolled bleeding from esophageal varices not responding to at least 2 sessions of sclerotherapy or banding
2. Bleeding gastric or ectopic varices
3. Hypersplenism or massive symptomatic splenomegaly
4. Budd-Chiari Syndrome (selected patients)
5. Lack of access to endoscopic treatment
6. Symptomatic biliary obstruction due to choledochal varices
7. Inability to follow up repeatedly for endoscopic therapy

TYPE OF SURGICAL PROCEDURES:

Surgical procedures for portal hypertension can be broadly classified into three groups:

1) Porta-Systemic Shunts
2) Non Shunt surgery – Devascularisation
3) Liver transplantation
PORTA-SYSTEMIC SHUNTS

The aim is to divert blood flow from portal system to systemic circulation by anastomosing the portal vein or its tributaries i.e. splenic vein or superior mesenteric vein to renal vein or IVC in order to reduce pressure in the varices.

Even when indicated all children cannot be subjected to shunt surgery. About 20% are unfit for a shunt surgery. The choice of surgical procedure depends on the pathological process and its effect on the child. These can be used in acute uncontrolled hemorrhage scenario or for chronic Extrahepatic Portal venous obstruction.

CLASSIFICATION

1) NON SELECTIVE SHUNTS
   • Total Shunts – Portacaval, Mesocaval, Proximal Splenorenal shunt.
   • Partial Shunts – Small diameter Porta caval(Sarfeh)

2) SELECTIVE SHUNTS – Distal splenorenal shunts

Total shunts:

The main characteristics of total shunts are:

1. These completely divert portal blood flow into the systemic circulation, hence are highly effective in controlling acute and long term variceal bleeding > 90%.
2. These shunts have a wider diameter, so chances of shunt thrombosis are rare.

The disadvantages with total shunts are:

1. There is no portal vein flow to the liver which results in diversion of hepatotropic factors leading to deterioration of liver functions, the other one being encephalopathy of up to 40% over the long term, which depends on the liver status of the child.
2. If graft is utilized, thrombosis of the shunt is frequent.
3. Ascites does not get relieved after the shunt surgery.

a) Portal caval shunt:

The two different surgical techniques in portacaval shunts are the end to side portacaval shunt as first described by Eck (2) and the side to side shunt. Portal blood is completely redirected into the inferior vena cava below the liver and the hepatic end of the portal vein is oversewn in the end to side shunt. It is the choice of shunt surgery in the emergency control of variceal bleeding where medical management has failed as it can be readily constructed (3).

The side to side portacaval shunt allows blood from the intestine and the spleen to flow easily into the IVC. In addition, the hepatic end of the portal vein is changed into an outflow tract thus decreasing sinusoidal hypertension. Thus it is the shunt of choice in Budd –Chiari post hepatic PHT, resulting in long term palliation of the disease with arrest or delay in the progression of hepatic fibrosis and ultimate failure (4). Other effective operations in this condition are mesocaval and central splenorenal shunts, which allow portal blood in the liver to empty in a retrograde fashion through the patent portal vein.
b) **Mesocaval shunt**: It is either constructed directly as a side to side anastomosis between the two veins or with the interposition of a short autologous vein graft - internal jugular vein or a prosthetic graft (5).

Vein graft is preferred as the incidence of thrombosis is particularly less. It has been used in children in a variety of settings and diseases with uniformly acceptable results (6). This is preferred by some as it allows an anastomosis between two large diameter vessels and because the length of the anastomosis can be increased to some extent to facilitate the creation of a large venous fistula. Splenectomy need not necessarily be done, except in cases with hypersplenism and thus preventing post splenectomy sepsis (7).

C) **Proximal Splenorenal Shunt (Linton)**: The Splenic vein is divided close to the spleen and the mesenteric vein is sutured to the side of the left renal vein so that all the blood from the SMV and the IMV is shunted into the systemic circulation through the left renal vein (8). It invariably includes splenectomy hence it is popular choice in our country particularly for patients with EHPVO with large symptomatic splenomegaly and Hypersplenism.

**Partial Shunts**:

Sarfeh created a small diameter shunt between the portal vein and the IVC using PTFE graft as a conduit thus enabling enough diversion of blood from the portal circulation, dropping pressure in the mesenteric vessel thus decompressing the varices and reducing the chance of bleeding while maintaining enough pressure in the portal bed to allow hepatopetal flow and hepatic blood flow preservation (9). One important drawback is the increased incidence of thrombosis and recurrence of bleeding (10).

**Selective shunts –Warren or Distal Splenorenal Shunt**

The various selective shunts described are:

1. The distal splenorenal shunt (Dean Warren shunt)
2. Inokuchi Splenocaval (IMV to IVC)
3. Interposition shunts with the left gastric vein to inferior vena cava

**Characteristics**:

1. Selective shunts decompress the varices only, and presumably only a part of the portal circulation is decompressed
2. Portal pressure and portal flow are not affected much.
3. Since the portal perfusion is maintained via the mesenteric supply it reduces the risk of postoperative hepatic encephalopathy, about 15%
4. These shunts produce ascites
5. It does not interfere with future liver transplant

**Disadvantages**
1. Since it produces ascites, it is contraindicated in those children with massive intractable ascites prior to the shunt procedure.
2. This shunt cannot be done in children who had undergone previous splenectomy due to any reason.

Warren (11) et al described a shunt between the portal end of the splenic vein and the side of the renal vein. The Splenic confluence with the portal vein is ligated and the coronary vein also ligated. In this manner the gastro esophageal varices were decompressed across the short gastric vessels and the spleen into the renal vein. Long term patency rates exceed 90% with resolution of bleeding in all patients with patent shunts.

Distal splenorenal shunting is used primarily in children with extra hepatic portal vein thrombosis, stable Child class A or B cirrhosis or less common forms of intrahepatic portal hypertension such as congenital hepatic fibrosis with well preserved liver function but symptomatic variceal bleeding. It has also shown a lot of benefit in children with advanced Hypersplenism (12).

**Mesenteric to left portal vein bypass (REX shunt)**

This is the only physiological shunt as it maintains the normal flow of blood through the liver. For those patients with cirrhosis with extra hepatic portal venous hypertension with well preserved liver function, definitive treatment may be required for recurrent bleeding or for progression of hypersplenism to the point that it interferes with the normal activity of the child (13). The treatment of EHPVO has been evolving since the advent of the Rex shunt in 1992. It was originally described to treat portal vein thrombosis after liver transplants. Deville Goyet (14) first described this technique in reconstituting portal venous outflow tract in non transplant patients. Essentially this operation relieves portal hypertension by redirecting the blood flow from the obstructed mesenteric system to the still patent intrahepatic portal vein. The prerequisites for a successful bypass operation includes:- absence of intrinsic liver disease, a patent intrahepatic portal tree and a suitable vein in the mesenteric circulation as a suitable inflow tract for mesenteric blood. Evaluation of the intrahepatic portal vein has been done by USG Doppler, Angiography, Direct portal venography and MRA. However Daniel et al (13) has described direct visualization of the intra hepatic portal vein as the best current method. The vein is located in the recessus of Rex. Autologous vein graft i.e. Internal jugular vein is used as a conduit between the intrahepatic portal vein and the mesenteric vein. The main advantage of this type of shunt is that it is restorative rather than palliative. It restores the portal flow to the liver and relieves the symptoms of PHT. Patency rates exceed 90% which depends on the quality of the intrahepatic portal vein and the number of branches that allows sufficient runoff.

**NON SHUNT SURGERY - DEVASCULARISATION**

The aim of devascularization procedures is direct disconnection between the portal and azygos vein. There are several variations in the operative procedures described by several authors. These devascularisation procedures, also known as portal non -decompressive procedures or porto-azygos disconnection include procedures which aim to control the gastro esophageal varices either by direct on varices(variceal ligation of esophageal varices or esophageal/gastric transection) or by disconnecting varices from their feeding vessels.
The main steps of the procedure as described by Siguira and Futagawa (15) involves transthoracic extensive devascularisation of lower esophagus from the level of the left inferior pulmonary vein up to the diaphragm, esophageal transaction followed by end to end anastomosis, transabdominal devascularisation of upper half of the stomach, splenectomy, vagotomy and pyloroplasty. A modified Siguira operation in children has been described in which the spleen is preserved and staple anastomosis of the esophagus is done (16). It was originally described as an emergency operation to control variceal bleeding, but it has now fallen into disrepute because of high incidence of variceal rebleeding (30-50%) (17).

**LIVER TRANSPLANTATION**

Irrespective of the etiology of portal hypertension, clinical end results are the same - bleeding from varices, ascites, splenomegaly with hypersplenism. Although major acute emphasis in the treatment of PHT has been control of bleeding, many children with intrinsic liver disease continue to lose hepatic functional reserve. Thus liver transplantation often becomes the reasonable therapeutic option for patients with advanced liver disease. (18)

**Our experience**

About 110 extrahepatic portal hypertension patients are registered in the Pediatric Liver Clinic at B.J.Wadia Hospital for Children. We have over 550 patients with liver disease registered in the clinic. We had analyzed data of 35 children with portal hypertension (both Extrahepatic and intrahepatic) in 2005-2006 of ages between 10 months to 10 years. The M:F ratio was found to be 1.7:1. Presenting complaints were variable, almost 90% presenting with hematemesis or malena or both. Associated complaints were jaundice in 18 children, ascites in 11, hepatomegaly in 16 and splenomegaly in 23 children of which 10 had hypersplenism. The cause of portal hypertension was evaluated and 15 of these had extrahepatic portal venous obstruction, 5 operated children with Biliary atresia, 5 with Wilson’s disease, 4 with chronic liver disease, 3 GSD, 2 Thalassemia major, and 1 child with congenital hepatic fibrosis. Majority of the children underwent control of hematemesis by endoscopy and sclerotherapy or banding of esophageal varices. Shunt surgery was performed in 8 children (22.9%). Two children underwent devascularization procedure of which one is having persistent hematemesis and malena. Two of the 35 children had associated Protein S deficiency and Anti-phospholipid syndrome.

**SUMMARY**

A child with portal hypertension presents a unique challenge to the surgeon. The number of treatment options has increased manifold ranging from new medications which reduce the pressure in the mesenteric tree to endoscopic sclerotherapy/banding. Endoscopic treatment is highly effective and appears to be the treatment of choice in the initial management of esophageal varices in children. Porta systemic shunts still has a role in the management of PHT and is reserved for indications like bleeding unresponsive to sclerotherapy, gastric or ectopic variceal bleeding, symptomatic massive splenomegaly or Hypersplenism, isolated EHPVO. It is the etiology of portal hypertension, particularly the presence or absence of intrinsic liver disease that dictates the short term and long term outcome. In patients with normal liver and extra hepatic obstruction, shunt is the definitive treatment. In patients with intrinsic liver disease, severity of bleeding dictates the management. In patients with stable liver function shunt
can be performed with 100% short term and 92% long term results. Liver transplantation is offered for shunt failures or patients with poor hepatic function.

REFERENCES

2) Eck: On the question of ligature of portal vein. Voyenno med 1973;130:1
6) Altman RP: Portal decompression by interposition mesocaval shunt in Biliary Atresia patients. JPS 1976;11:809-14
17) Wexler: Non shunting operations for variceal hemorrhage. Surg Cl No Am.70;426:1990