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Role of Shunt Surgery in Non-Cirrhotic Portal Hypertension- A Case Series

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Abstract

Background: Surgical treatment of NCPH (non-cirrhotic portal hypertension is less commonly performed nowadays due to advances in endoscopic and interventional radiological methods. The aim of this study was to evaluate the efficacy and safety of surgical porto-systemic shunting as a one-time treatment of non-cirrhotic portal hypertension (NCPH).

Methods and Materials: 79 patients with NCPH; 35 Extra Hepatic Portal Vein Obstruction (EHPVO), 33 Non-cirrhotic Portal Fibrosis (NCPF), 4 Congenital Hepatic Fibrosis (CHF), 3 chronic Budd Chiari syndrome (BCS), 2 Schistosomiasis, 1 agenesis of Right hepatic Vein (RHV) and another with hypoplasia of right hepatic lobe; underwent some porto-systemic shunt surgery with or without splenectomy. 1 patient died due to pulmonary embolism after surgery for Budd Chiari Syndrome. 3 patients had developed shunt thrombosis (2 synthetic PTFE grafts) at 12 months and one developed shunt thrombosis after 24 months. Tere was no rebleed in any patient and liver function remained normal in all patients.

Discussion and Conclusion: Porto-systemic shunt surgery is safe and effective in the management of non-cirrhotic portal hypertension and should be encouraged as a one-time treatment. Shunt surgery offers a one-time treatment for NCPH and avoids prolonged and repeated follow up as compared to endoscopic management alone.

Keywords: Shunt Surgery; Non-Cirrhotic Portal Hypertension; Liver function tests; Esophago-gastric devascularization; Variceal haemorrhage

Abbreviations: NCPH: Non-Cirrhotic Portal Hypertension; EHPVO: Extra Hepatic Portal Vein Obstruction; NCPF: Non-cirrhotic Portal Fibrosis; BCS: Budd Chiari syndrome; RHV: Right hepatic Vein; LFT: Liver Function Tests; CECT: Contrast Enhanced Triphasic Computerized Tomography; ALKP: Alkaline Phosphatase; GGT: Gamma Glutamyl Transferase; PHB: Portal Biliopathy; IVC: Inferior Vena Cava; TIPS: Trans-Jugular Intrahepatic Porto-Systemic; PHG: Portal Hypertensive Gastropathy; PSS: Porto-systemic Shunt Surgery; EGD: Esophago-Gastric Devascularization; PTFE: Polytetrafluorethylene; PSRS: Proximal splenorenal Shunt; IPH: Idiopathic Portal Hypertension; ADPKD: Autosomal Dominant Polycystic Kidney Disease; ARPKD: Autosomal Recessive Polycystic Kidney Disease

Introduction

Liver cirrhosis is the commonest cause of portal hypertension, accounting for > 90% cases in the developed world and around 80% in developing countries in Asia and Africa [1]. Portal hypertension in patients without cirrhosis can be classified based on the site of resistance to blood flow as "prehepatic," "hepatic," and "post-hepatic." The "hepatic" causes of NCPH can be subdivided into "presinusoidal," "sinusoidal," and "post-sinusoidal" [2]. Non cirrhotic portal hypertension causes significant morbidity from recurrent variceal bleeding, hypersplenism, growth failure, portal biliopathy and ectopic varices [3,4]. Porto-systemic shunt surgery is an attractive option with one time treatment with long term durable effects in preventing variceal bleeding and relief from other symptoms of NCPH. The numbers seemed to have reduced with the recent advances in interventional radiology, and availability of endoscopic facilities. The aim of this study was to evaluate the safety and efficacy of shunt surgery in this patient population to assess its relevance in today's time and age.

Methods and Materials

79 patients with non-cirrhotic portal hypertension were operated between January 2013 to December 2022. All relevant data was prospectively collected. Written informed consent was taken from all patients for collection of data and use of the data for clinical analysis. All patients were followed up for a between 12 months to 108 months. Relevant investigations were done to assess liver function, bone marrow function, structural integrity of the liver and vascular anatomy of the portal venous system. Blood tests included liver function tests (LFT), complete hemogram, and renal function tests. Relevant investigations were done to assess liver disease and function. All patients underwent esophago-gastro-duodenoscopy before surgery, and at 6 months, 12 months

and at 24 months. All patients underwent contrast enhanced triphasic computerized tomography (CECT) and the liver was evaluated for size, surface nodularity, vascular inflow and patency of the portal circulation including splenic and superior mesenteric veins. Ultrasound abdomen was done to look for free fluid and any biliary calculi. In patients with conjugated hyperbilirubinemia, raised alkaline phosphatase and gamma glutamyl transferase (ALKP and GGT) an MRI (magnetic resonance imaging) with MRCP (magnetic resonance cholangio-pancreatography) was done. All patients underwent a bone marrow examination in view of pancytopenia and possibility of myeloproliferative disorder, which would preclude a splenectomy. Patients planned for splenectomy received pre-splenectomy vaccination at least one week before surgery. Vaccination for Hemophilus influenza, Pneumococcus and meningococcus was done.

Findings

The aetiology of non-cirrhotic portal hypertension was as follows: (Table 1). All patient shad a history of variceal bleeding and were referred for management of portal hypertension, while some patients were referred for liver transplant evaluation. Surprisingly 6 patients, suffering from EHPVO, between the ages of 50y and 60y were found to have grade 1 and grade 2 myeloproliferative disorder (myelodysplastic syndrome) which were excluded from our study. There were 35 patients with EHPVO (extra-hepatic portal vein obstruction), 33 patients with NCPF (non-cirrhotic portal fibrosis), 3 patients with chronic Budd Chiari syndrome, 2 patients of schistosomiasis, one patient with complete agenesis of right hepatic vein and another with hypoplasia of right hepatic vein. 15 patients had some evidence of portal biliopathy (PHB). 9 patients had obstructive jaundice, of which 2 were due to choledocholithiasis, 2 had haemobilia and required endoscopic covered metal stent placement, 2 patients had extrinsic compression of the common bile duct by the portal cavernoma.

Table 1: The aetiology of non-cirrhotic portal hypertension.

NCPF	33
EHPVO	35
Congenital hepatic fibrosis (CHF)	4
Budd Chiari Syndrome (Chronic)	3
Schistosomiasis	2
Agenesis of Right hepatic vein	1
Hypoplasia/ Aplasia of right Hepatic Vv.	1

68 patients underwent a splenectomy with PSRS (proximal spleno-renal shunt (Linton Shunt)) (Figure 1, 2). 1 patient underwent central spleno-renal shunting, involving side to side anastomosis of the splenic and renal vein (Figure 3). 2 patents underwent a superior mesenterico-common iliac shunt; 4 patients, one Budd Chiari syndrome and another previous splenectomy with spleno-portal thrombosis, and 2 patients with NCPF; underwent a meso-caval shunt (ringed PTFE graft) (Figure 4), 1 patient with Budd Chiari syndrome received a porto-caval shunt (PTFE graft); and 1 underwent shunting between the inferior mesenteric vein and the renal vein. Another patient underwent an inferior mesenteric vein - IVC (inferior vena cava shunt), both of which indirectly becomes a splenorenal shunt (PSRS). One patient with Budd Chiari (BCS) underwent a meso-cavo-atrial shunt. If synthetic PTFE graft was used, all anastomoses were done with 6-0 prolene continuous sutures after priming the graft with heparinized saline. If synthetic graft was not used, all anastomoses were done with 5-0 prolene, continuous sutures for the posterior wall and interrupted sutures for the anterior wall. The splenic artery was first ligated and the spleen was allowed to decongest, before the splenectomy.

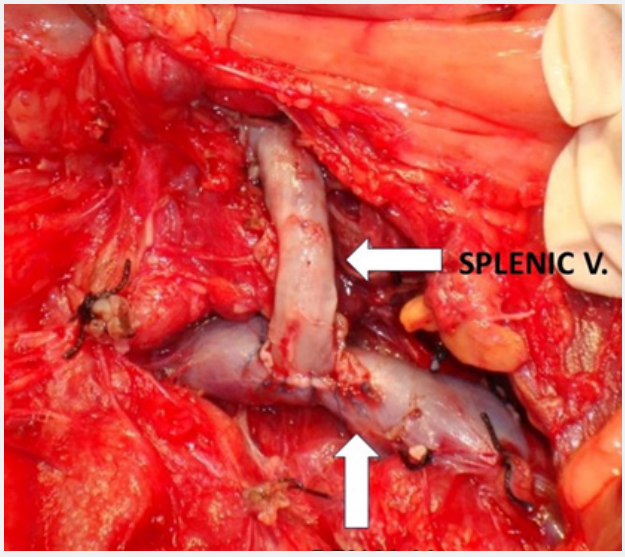

Figure 1: Standard PSRS (proximal spleno-renal Shunt (Linton Shunt)

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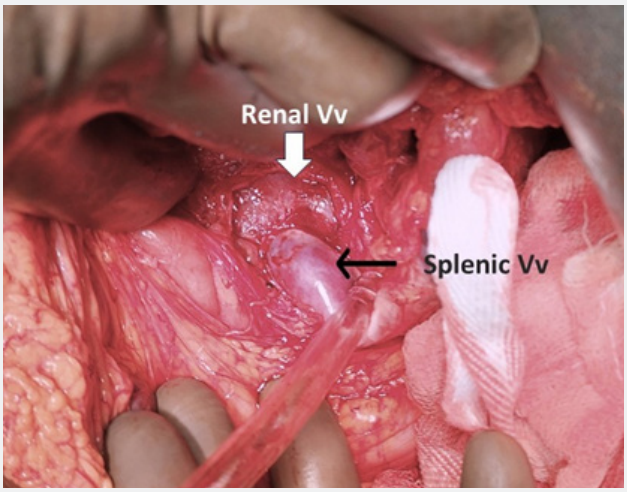

Figure 2: PSRS in a patient with a long splenic vein.

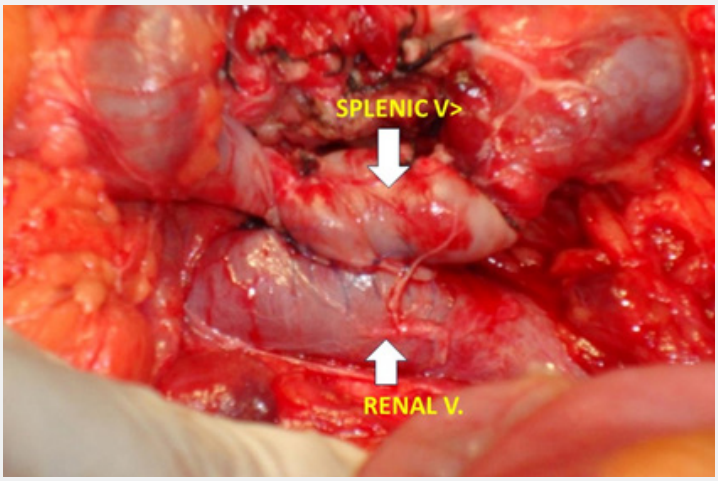

Figure 3: Central Spleno-renal Shunt (side to side spleno-renal shunt).

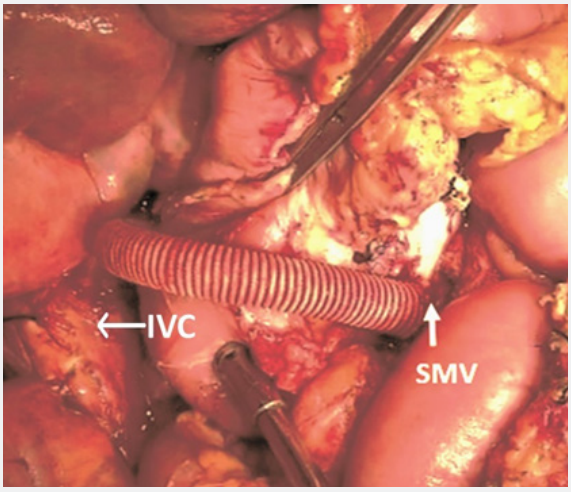

Figure 4: Meso-Caval Shunt using a PTFE graft.

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Total blood loss during surgery ranged from 180ml to 900ml (mean - 420.37ml) including the blood contained in the spleen of splenectomized patients. 6 patients required transfusion of packed RBC (red blood cells) and one patient was given platelet transfusion, who had a precipitous fall in platelet count due to heparin induced thrombocytopenia. All patients were given 3000U of unfractionated heparin intravenously at the time of the vascular anastomosis. Post operatively, unfractionated heparin was given in prophylactic dose at 5000IU q8hrly sub-cutaneous till oral intake was resumed. Thereafter anti-platelet in the form of aspirin 75mg per day was commenced and continued for 6 months in patients without prosthetic grafts. Patients with PTFE grafts were anticoagulated with warfarin titrated to an INR between 2 to 2.5, and warfarin was continued for life. Hospital stay for all patients ranged from 6 days to 9 days.

2 patients had had previous splenectomy, 1 patient as part of a devascularisation procedure 15 years ago, while another patient had a splenectomy in childhood for splenic trauma. All the remaining patients had some degree of hypersplenism. 1 patient with Budd Chiari syndrome died in the first week post-surgery secondary to pulmonary embolism. He had undergone a meso-cavo-atrial shunt. Another patient died secondary to covid illness 1 year after having undergone a splenectomy with a proximal splenorenal shunt for non-cirrhotic portal fibrosis. 4 patients with schistosomiasis, 1 patient suffering from EHPVO, 1 from Congenital Hepatic Fibrosis and 1 patient with NCPF did not have a shuntable vein, and underwent esophago-gastric devascularization with splenectomy. These were not included in the present study.

Outcomes

3 patients developed shunt thrombosis at 12 months from the surgery and one patient with EHPVO was found to have a thrombosed shunt at 2 years after surgery. One patient with EHPVO was found to have small bowel stricture during surgery, with adhesions and a site of entero-enteric fistulation. Histological examination of the resected small bowel showed Crohn's disease, and the patient is being managed medically for the same. The same patient developed a thrombotic stroke 4 years from the surgery, but a coagulation work up turned up negative for all pro thrombotic conditions. The patient has been on anti-coagulants since then and has made am almost complete neurological recovery. Another patient, an elderly female, developed a sigmoid colon cancer 5 years from the porto-systemic shunt surgery, and was operated for the same. She has been on adjuvant chemotherapy, and the liver function remains normal. All patients had protocol endoscopy at 12 months and at 24 months. No patient had any varices and there was no incidence of variceal bleeding in any patient.

Two patients with NCPF, went on to develop autoimmune liver disease 4 and 6 years after surgery. One of these patients was detected with cirrhosis at 6y of follow-up. Both the patients are now being managed medically, and the liver function remains normal now. The patient who has developed autoimmune liver cirrhosis, had transiently decompensated with development of ascites, transaminitis and low albumin levels, However, with initiation of therapy the liver function returned to normal and remains symptom free as of now. One patient travelled to his home country and only blood tests were available for follow up. In 14 patients, portal biliopathy resolved with porto-systemic shunting, while one patient still has a biliary stent in situ with normal liver function. Since all patients did not have an MRI examination, the actual incidence of portal biliopathy is difficult to ascertain.

Discussion

Rosemurgy pointed out that, "non-surgeons increasingly control the care of patients experiencing variceal bleeding. This has promoted the application of nonsurgical therapies, like trans-jugular intrahepatic porto-systemic stent shunt (TIPS). Therefore, surgeons-in-practice are generally not involved, and surgeons-in-training see fewer patients with variceal bleeding than in days past. Thereby, most surgeons believe that porto-systemic shunting is not in their therapeutic armamentarium, further promoting the involvement of non-surgeons and the application of TIPS" [5]. The art of surgery for porto-systemic shunting for portal hypertension seems to be dwindling with lesser experience and lack of training. Decline in the skills required for porto-systemic shunt surgery has happened over time with reduction in the involvement of surgeons in the care of patients with portal hypertension [6]. TIPS is now more widely available and even the Baveno VI consensus statement recommends TIPS if primary therapy with pharmacotherapy and endoscopic variceal ligation fails in control of bleeding [7]. Common clinical features of NCPH include upper gastrointestinal bleeding; hypersplenism and severe anaemia with resultant effects of anemia and malnutrition; portal biliopathy; and portal hypertensive gastro-enteropathy with resultant malnutrition; especially in children [8,9]. The aim of treatment in NCPH is reduction of portal pressure to prevent complications of portal hypertension; treatment of hypersplenism.

Primary prophylaxis of variceal haemorrhage is probably difficult, as the most common presentation of non-cirrhotic portal hypertension is upper gastro-intestinal haemorrhage from esophageal varices [2]. Treatment of acute variceal haemorrhage can usually be accomplished by endotherapy with pharmacotherapy [10]. Small fraction of patients may end up needing emergency surgery for acute variceal bleeding [11]. In the acute setting or as secondary prophylaxis; initial experience was with esophago-gastric devascularization with esophageal transection. Esophago-gastric devascularization without esophageal transection is equally effective with lesser morbidity [12,13]. Endoscopic therapy and secondary prophylaxis required repeated sessions and prolonged follow up. Over a prolonged period of follow up, surgery and endotherapy are equally effective with slightly more rebleeds after endotherapy (17% for endoscopic treatment vs 11% after surgery) [14,15]. Even though long-term rebleeding rates with endotherapy and surgery are similar, endotherapy requires stringent follow up, prolonged

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treatment, and repeated sessions; which may lead to esophageal strictures, ectopic varices and congestive gastropathy (Portal Hypertensive Gastropathy - PHG). Endotherapy however, does not seem to solve the problems of hypersplenism, growth retardation, portal biliopathy and recurrent pain from infarcts in fairly massive splenomegaly [7].

Surgery in NCPH aims to bypass the site of obstruction to the flow in the portal system; either through porto-portal bypass, like the Rex shunt; or more commonly with porto-systemic shunting. When shunting or bypass is not feasible other surgical options like devascularisation with or without splenectomy resulting in disconnection of the sites of bleeding from the portal circulation. Surgery is indicated in patients who are symptomatic and have had multiple episodes of gastro-intestinal bleeding; in those with failure to thrive and nutritional ill effects of portal hypertension. Patients with symptomatic hypersplenism form another group of patients likely to benefit most from surgery. Surgery also needs to be considered in patients who have limited access to healthcare, those who are likely to travel to remote places; and those who desire a single stage correction of portal hypertension. In this group of patients with non-cirrhotic portal hypertension, liver function is very well preserved and hepatic encephalopathy did not seem to occur even after non-selective shunts when studied by other groups [16,17]. Surgical options in these patients can be divided into 3; porto-systemic shunt surgery (PSS), esophago-gastric devascularization (EGD) and the recently described Rex shunt [porto-portal or (mesenterico-left portal vein) bypass]. Shunt surgery remains a one-time treatment procedure with durable, long-term efficacy in preventing variceal rebleeding, and preventing morbidities associated with NCPH [18].

Surgical shunting may be selective or non-selective; and partial or complete.

Selective shunts: Selective shunting involves diversion of a part of the portal circulation away from the portal vein to reduce the portal inflow, maintaining some portal flow to the liver [18].

i. Distal splenorenal shunt: This shunt intervention connects the distal splenic vein to the left renal vein, with or without spleno-pancreatic and -gastric connection.

ii. Inokuchi shunt: Shunting of the coronary vein to the inferior vena cava (IVC) to divert blood from the stomach and oesophagus relieves the left sided portal hypertension which has a high risk of bleeding varices [19].

Non-selective shunts: partial/ total

Partial shunts: Small-diameter H-graft shunt. This shunt is made with polytetrafluorethylene (PTFE)reinforced grafts that measure 8 mm in internal diameter and connects the superior mesenteric vein or portal vein to inferior vena cava. Small-diameter TIPS (Trans-jugular intra-hepatic porto-systemic shunt). This is TIPS constructed with stents measuring 8 mm or less in internal diameter, or primarily constrained to reduce the internal diameter.

Complete/ Total shunts: In these types of shunts, no effort is made to maintain any hepatopetal flow, and all flow may be diverted away from the liver. The common total shunts are portocaval shunts (either side-to-side or end-to-side) (Orloff 2007); central splenorenal shunts (constructed by anastomosing proximal splenic vein to left renal vein); and the large diameter H-graft shunts constructed with 16 mm polytetrafluoroethylene (PTFE) prosthesis (Sarfeh 1986) [20].

i. Portocaval shunt: This shunt is constructed to connect the portal vein to the inferior vena cava directly, which could be end to side after division of the portal vein or side to side using a graft or autologous conduit.

ii. Mesocaval shunt: This shunt is constructed to connect the superior mesenteric vein to the inferior vena cava directly, often using a prosthetic graft.

iii. Central (proximal) splenorenal shunt. This shunt is constructed to connect the proximal splenic vein to the left renal vein, with or without spleno-pancreatic and -gastric disconnection or splenectomy.

iv. Large-diameter H-graft shunt: This shunt is constructed with polytetrafluorethylene reinforced grafts that measure 16 mm or greater in internal diameter and connects superior mesenteric vein or portal vein to the inferior vena cava.

v. Clatworthy shunt: End of the superior mesenteric vein (SMV) is connected to side of the common Iliac vein, enabling the reversal of the portal flow into the Iliac vein [21].

vi. The diameter of the shunt controls the diversion of flow, and a shunt diameter of more than 8mm is supposed to be adequate at compete diversion of the portal flow to the systemic circulation [22]. Non-conventional shunt surgery, using un-named portal venous radicals anastomosed to systemic veins, are effective in patients unsuited for other shunts, especially PSRS, and it achieves the desired effects in a significant proportion of patients [23].

vii. Rex shunt: The Rex shunt is a superior mesenterico-left portal vein bypass, typically useful in only patients with chronic EHPVO (extra-hepatic portal vein obstruction [24]. The shunt is usually done using an autologous vein graft (internal jugular or saphenous vein) to restore blood flow from the superior mesenteric vein to the left portal vein in the portal fissure in the recess of Rex.

viii. Proximal splenorenal shunt (PSRS) is the commonly used procedure in Indian center's for treatment of non-cirrhotic portal hypertension, especially in children and in patients with a large spleen with hypersplenism. PSRS effectively reverses hypersplenism due to the splenectomy and provides relief from the recurrent pain of splenic infarcts. Splenectomy also results in a reduced inflow to the portal flow thereby reducing the portal flow,

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and hence the portal pressure.

Removal of the spleen does not seem to offer any disadvantage in these patients as the incidence of overwhelming post splenectomy sepsis is very rare among this group of patients [19,6]. Good long-term patency rated of more than 90-95% have been reported by many studies [17].

Review of Literature

NCPF (non-cirrhotic portal fibrosis) and EHPVO (extra-hepatic portal vein obstruction) for the major causes of NCPH in this group.

NCPF

The pathogenesis of NCPH revolves around factors that cause portal venous attenuation at the small vessel level, subsequent phlebosclerosis and aberrant vasculature [2]. It is referred to as noncirrhotic portal fibrosis (NCPF) in south Asia, while in Japan and east Asian countries idiopathic portal hypertension (IPH). In the western world it is far less common, and called as hepatoportal sclerosis, noncirrhotic intrahepatic portal hypertension, and idiopathic noncirrhotic intrahepatic portal hypertension [25,26]. Portal hypertensive bleed which is well tolerated and pancytopenia are common presenting features in NCPF, though in rare instances late decompensation has been seen [27,29]. 5 percent of explant livers from patients undergoing liver transplantation for cryptogenic liver cirrhosis showed features histologically compatible with NCPF [28]

The outcomes of shunt surgery in patients with NCPF are generally good, with very good long-term patency and very low rebleeding rates (<3-19%); but a small incidence of encephalopathy, and a small incidence of complications like hepatopulmonary syndrome, glomerulopathy and myelopathy [13,29,30]. It is thought that NCPF may progress liver disease (ESLD) due to progressive reduction in blood supply to the periphery of the liver [31]. Aaron et all identified some genetic factors that could play a significant role, and yet secondary insults like infections and toxins may impact the presentation of these genes [32,33]. Some autoimmune abnormalities have been reported in patients with NCPF, including connective tissue disorders, autoimmune anemia, ankylosing spondylitis, and scleroderma [34-37]. Thus, patients being offered shunt surgery for NCPF should be carefully evaluated and patient education is important.

EHPVO

The Rex shunt or superior mesenterico-left portal vein bypass has the advantage of restoring the hepatic flow of the splanchnic blood. In our experience we did not encounter any patent left portal vein, making the Rex bypass unsuitable. Guerin et al found the Rex bypass possible in only 58% of their patients, and a further 25% failed during surgery [38]. The available comparisons between the Rex shunt and non-selective porto-systemic shunt failed to show any advantage of the Rex shunt [20,39]. The liver in EHPVO is normal in structure and function and correction of portal hypertension seems to restore a normal quality of life. Prophylactic shunt surgery may be offered to patients who have never bled, but have severe hypersplenism with other symptoms like, pain from the enlarged spleen, portal biliopathy, portal hypertensive gastro-enteropathy and growth retardation [40].

Schistosomiasis

Hepatosplenic schistosomiasis is caused by the lodged Schistosoma eggs in the liver parenchyma, which elicit a host immune response, provoking granuloma formation and peri-portal fibrosis [41]. As the disease progresses, collagen deposition and fibrosis may cause organ damage that may be only partially reversible.

Parasite burden, egg load, T-cell responses, infection in younger age and degree and duration of infection determine the course of disease. Praziquantel results in improvement in intestinal symptoms and inflammation improves rapidly, while liver pathology is only partially reversed [42]. Nordmann et al concluded that TIPS (trans jugular intra-hepatic porto-systemic shunt) and shunt surgery as safe and effective treatment for patients with portal hypertension due to Schistosomiasis, but requires careful patient selection [43].

Congenital Hepatic Fibrosis:

Liver biopsy remains the gold standard in diagnosis of CHF. CHF may have variable clinical features, ranging from asymptomatic to decompensated liver disease [44,45]. The age of presentation is also quite variable, from early childhood to 5th and 6th decade of life; but most commonly presents in adolescent children. It seems to be a ductal plate malformation, often associated with Caroli's disease, von Meyenburg complexes, autosomal dominant polycystic kidney disease (ADPKD), or autosomal recessive polycystic kidney disease (ARPKD) [46]. The most common presentations in these patients are splenomegaly, esophageal varices, and gastrointestinal bleeding due to portal hypertension. Based on clinical presentation, there are four clinical forms of CHF: portal hypertensive, cholangitic, mixed, and latent [47]. In portal hypertensive type, variceal bleeding seems to be the most disabling problem, and it has been observed that splenectomy combined with portosystemic shunting is a better choice for treating repeated variceal bleeding [48-50].

Agenesis/ Hypoplasia of Right Hepatic VEIN (RHV)

Agenesis of the right hepatic lobe is a very rare clinical entity and portal hypertension seems to accompany only a part of these [51]. Radin et al reported in 24 cases, that this anomaly may accompany with biliary tract disease, portal hypertension of the presinusoidal type, cholelithiasis and other congenital anomalies [52]. Liang et al, in 2017, in a study involving 41 patients with right hepatic lobe agenesis or hypoplasia; showed right lobe volume reduction and stenosis of the right portal vein in 100, ectopic gallbladder 95%, left lobe hypertrophy in 95%,

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left portal vein thickening in 90%, dextral displacement of the first hepatic portal in 78% cases, caudate lobe hypertrophy in 39% and portal hypertension in 34% patients [53]. Hypoplasia or agenesis of the right hepatic lobe is due to hypoplasia or agenesis of the right hepatic vein. It is associated with portal hypertension of the intrahepatic presinusoidal type [54]. Crowding of intrahepatic portal vein branches is not completely compensated by the hypertrophied left lobar portal vein branches, resulting in increased resistance in portal vein and thus portal hypertension [55]. Another hypothesis for portal hypertension is a potential shunt between the hepatic artery and the portal vein [57].

One patient had a complete absence of the RHV, with a normal liver biopsy, a large spleen with hypersplenism; and variceal bleed. Since portal hypertension was the only demonstrated anomaly, a surgical shunt with splenectomy was offered. The other patient had a very narrow right inferior hepatic vein, 2.3mm diameter. Since the liver biopsy was normal, and maintained normal liver function, hypoplasia of the right lobe of liver was considered and surgical shunting was offered. Both patients did dramatically well and remain symptom free.

Budd Chiari Syndrome

The condition may be primary, which is quite rare, while secondary Budd Chiari syndrome is commoner with a multitude of causes [56]. Clinically BCS may present as an acute, chronic or more commonly as acute on chronic. The clinical features may point towards a congested liver, with acute abdominal pain, enlarged liver and fever. Sometimes there may be features of liver cell failure with jaundice and ascites [57]. Patients with frank cirrhosis and decompensated liver disease are best treated by liver transplantation. Initial treatment involves anti-coagulation, but as the disease progresses, treatment is directed at decompressing the liver and reducing hepatic congestion; by converting the portal vein to an outflow tract [58], thereby facilitating hepatic arterial perfusion. The commonest method of achieving this is via a trans-jugular portocaval shunt (TIPS). Another option is surgical porto-systemic shunting, which though quite effective, seems to have lost favour due to reported high morbidity and mortality.

Though immediate success of TIPS procedure is more than 90% [58], the incidence of shunt dysfunction due to thrombosis, stenosis and intimal overgrowth is significant. The incidence of shunt dysfunction has reduced significantly with the advent of PTFE covered shunts as against bare shunts, but it remains significant. Early in 2004 Hernadez-Guerra et all found the actuarial rates of primary patency in the bare-stent group were 19% at 1 year compared with 67% in the PTFE-covered stent group [59]. A 2008 study found that the primary patency rate at 2 years was 12% using bare stents and 56% using covered stents [60]. Primary patency of covered TIPS was 66% after 1 year and 42% after 2 years, and after intervention it improved at 1 and 2 years to 83% and 79%, respectively [61]. This is leads to an increase in morbidity and costs of the procedure. In our practice, most patients are self-funded and the cost of a TIPS procedure usually offsets the cost of surgery. The advantages that TIPS provides, however, are significant; and hence TIPS has by and far replaced surgical intervention across the world.

TIPS is less invasive; the compression or occlusion of the IVC does not hinder TIPS placement. Surgical porto-caval or meso-caval shunts alone are inadequate due to hypertrophy of the caudate lobe with subsequent IVC compression, and additional cavo-atrial anastomosis is required . The TIPS, however, bypasses the stenosed IVC segment and operates with or without the enlargement of the caudate lobe [62].

However, due to the drawbacks of TIPS procedure, surgical intervention finds a place in the management of Budd Chiari syndrome. In carefully selected patients, Behera et al were able to show long term patency at a mean follow-up of 40 months with complete resolution of symptoms in a group of 10 patients [63]. Guang Chang et al in 2017, showed excellent long-term outcomes with meso-atrial shunting for treatment of Budd Chiari syndrome [64]. Orloff et al, have shown excellent outcomes with reversal of histological changes of BCS in patients treated with side-side portocaval shunt for hepatic venous occlusion and side to side porto-caval shunt combined with cavo-atrial shunt in the presence of IVC compression or blockage. They have shown 95% to 100% survival in these patients 5-38 years and 5-25years after surgery respectively [65].

Conclusion

Surgical porto-systemic shunts, with or without splenectomy, are a useful, safe and effective tool for treatment of non-cirrhotic portal hypertension. In patients with EHPVO, it should probably be the treatment of choice. In patients with EHPVO, if a Rex shunt is feasible, it may be attempted, but very commonly a Rex shunt is not possible a hypersplenism is common. Hence splenectomy with PSS should be offered as a one-time curative option in these patients. In NCPF, with careful selection of patients with patient education, one may be able to offer porto systemic shunt surgery for patients who have poor access to healthcare; who are residing at remote location and when variceal bleeding and hypersplenism affect the quality of life. However, since NCPF is a poorly understood condition, this needs to be investigated. In patients with schistosomiasis, Budd Chiari syndrome, congenital hepatic fibrosis, if portal hypertension is cause for the symptoms, then a surgical shunt may be offered with good outcomes. The treatment of non-cirrhotic portal hypertension should probably involve a multi-disciplinary approach, with well-trained surgeons and radiologists along with gastroenterologists and endoscopists. Lastly, surgical training in treatment of portal hypertension should not be neglected; as it is a very useful option and with very satisfying outcomes.

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