Current Role of Portosystemic Shunt Surgery in the Management of Hepatic Venous Outflow Obstruction

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Key Words
Budd-Chiari syndrome \cdot Hepatic vein thrombosis \cdot Shunts \cdot Portosystemic shunts \cdot Transjugular intrahepatic portosystemic shunt \cdot Angioplasty

Abstract

Background: Hepatic venous outflow obstruction (HVOO) is a rare disorder that occurs predominantly due to a hypercoagulable state. The syndrome may result from hepatic vein obstruction, inferior vena cava obstruction or a combination of both and manifests with post-sinusoidal portal hypertension. The presentation may be fulminant with poor prognosis or as either acute, subacute or chronic forms with relatively better prognosis. Portosystemic shunt surgery (PSS) has thus far been the mainstay of treatment for HVOO. However, over the last decade, transjugular intrahepatic portosystemic shunt (TIPS) and liver transplantation have emerged as viable options. This review aims to evaluate the available treatment options and the current relevance of PSS for the management of HVOO. Methods: A literature review on investigations and treatment was performed using Medline and additional library searches. Results: Portosystemic shunts form the mainstay of treatment for patients with subacute presentation (preserved liver function) with medically intractable ascites or recurrent variceal hemorrhage. Excellent results with 5-year survival of more than 90% have been reported from specialized centers. The main limitation for PSS is the reported perioperative mortality of 10–20% and a declining technical expertise for such surgery. Liver transplantation with disease-specific 5-year survival between 50 and 95% is presently the treatment of choice for patients with fulminant presentation, end-stage liver disease (ESLD), unshuntable anatomy or decompensation after PSS. TIPS may be preferable for sick patients with acute presentation with isolated hepatic vein thrombosis or as a temporizing measure for those with ESLD awaiting transplantation. The drawback of TIPS is late shunt dysfunction that occurs in more than 50% of patients at 1 year. Conclusions: Due to rarity of the disorder there is a lack of trials comparing the different treatment modalities. Hence, the current treatment recommendations are based on retrospective studies. In a select group of HVOO patients (subacute presentation with preserved liver function), PSS remains the treatment of choice with excellent long-term results.

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Introduction

Hepatic venous outflow obstruction (HVOO) is a rare syndrome that results from blockage of the venous drainage of the liver due to hepatic vein thrombosis and/or inferior vena cava (IVC) obstruction. The clinical manifes-
tations range from fulminant hepatic failure to those of portal hypertension (ascites or variceal hemorrhage) or cirrhosis with end-stage liver disease (ESLD).

The last decade has seen rapid advancements in the investigations and treatment of HVOO. A cohort study of 120 patients managed between 1970 and 1992 reported that there was a statistically significant difference in 1-, 5- and 10-year survival between patients managed before and after 1985. This improvement was attributed to better management of hypercoagulable states and rising standards of general care [1].

Until the 1990s, portosystemic shunt surgery (PSS) with 5-year survival ranging between 57 and 95% formed the mainstay of the management [2–5]. The main limitations of shunt surgery are declining expertise and perioperative mortality of 10–20% [3, 4, 6, 7]. The predominant role of PSS has been challenged by the recent emergence of interventional radiology and strides made in liver transplantation. An impressive 5-year survival of 74% following transjugular intrahepatic portosystemic shunt (TIPS) for HVOO has recently been published [8]. Also, a 5-year survival of up to 95% has been reported for liver transplantation for HVOO [9].

This article presents an overview of the investigations and treatment options for the management of HVOO and evaluates the place of PSS in the current management algorithm.

**Terminology**

The Budd-Chiari syndrome (BCS) was first described by George Budd [10] in 1844 at King’s College Hospital, London. The clinical syndrome was characterized by Hans Chiari in 1899. Parker [11] coined the term ‘Budd-Chiari syndrome’ and defined the syndrome as ‘symptomatic occlusion of hepatic veins’ [12].

While hepatic vein occlusion was the more common cause of this syndrome in the West, obstruction of the hepatic portion of the IVC was the cause of BCS in Asia and South Africa. By definition, BCS was limited to hepatic vein occlusion and therefore an alternative terminology was proposed ‘hepatic venous outflow obstruction’ to include the obstructive lesions of the IVC [12, 13]. Okuda et al. [12] argued that the classical BCS and IVC obstruction have distinctive epidemiological and clinical features and suggested the term ‘Obliterative hepato-cavopathy’ to distinguish the IVC lesions from hepatic vein lesions.

A group of European experts has recently ‘redefined’ BCS as HVOO at any level from the small hepatic veins to the junction of the IVC and the right atrium regardless of the cause of obstruction. The outflow obstruction caused by hepatic veno-occlusive disease has been excluded from this definition. The authors argue that this term has withstood the passage of time and is more concise than any other proposed alternative [14]. However, the precise terminology awaits consensus between the experts of the East and the West.

The current review uses the term HVOO only because of anatomical considerations.

**Epidemiology**

HVOO is a rare cause of portal hypertension. In one of the early studies from Japan, the incidence was 4.9% of all portal hypertension patients [12, 15]. In India, the corresponding figure is 7–9% [6]. In recent articles from the western centers, this data is lacking [4, 5, 9, 14, 16, 17]. According to a questionnaire survey in France, the incidence is 0.36 per million per year corresponding to 20 new patients every year [18].

There are important differences in the syndrome between Asia and Africa on the one hand and the West on the other. Hepatic vein obstruction is the predominant lesion in the West while IVC obstruction is the more common lesion in Asia and Africa. The latter follows a more indolent clinical course and may be more frequently associated with liver cancer [6, 12, 17]. Amongst the Asian countries, there are differences in the etiology of the IVC obstruction. The etiology of HVOO is membranous obstruction of vena cava (MOV C) in Japan and Nepal while in India IVC thrombosis is more frequent [6, 12]. A combination of hepatic vein and IVC obstruction is common in India while in the Netherlands the syndrome of hepatic venous obstruction predominates [6, 19, 20].

**Etiology**

The major etiological factors responsible for HVOO include hypercoagulable states, membranous obstruction of the IVC, pregnancy and oral contraceptives, tumors and inflammatory conditions.

Hypercoagulable States
One of the common prothrombotic states in Europe is myeloproliferative disorder of which polycythemia vera is the most frequent. In the Netherlands and France, this classic disorder has been detected in 20–30% patients [21,
If the occult forms of this disorder are also considered, its prevalence rises to approximately 50% [17]. In Indian studies, myeloproliferative disorder occurs in less than 5% patients [19, 23].

The other common prothrombotic disorder is factor V Leiden (aberrant factor V resistant to destruction by activated protein C) identified in 25–30% patients [21, 24, 25].

The uncommon hypercoagulable disorders include protein C and S deficiency (more common than the former), antithrombin III deficiency (suggested by refractoriness to anticoagulant action of heparin) and antiphospholipid syndrome (young patients with arterial thrombosis, women with recurrent loss of pregnancy or thrombosis in abnormal locations) [26]. The presence of multiple disorders in the same patient is being increasingly recognized [27]. Other rare causes of HVOO include paroxysmal nocturnal hemoglobinuria and Bechet’s syndrome.

**Obstruction of the IVC**

This may take either of the two forms – membranous obstruction and IVC thrombosis.

**MOVC.** This is a major cause of HVOO in India, Japan and Nepal. In India, the incidence ranges between 26 and 28% while it is still higher in Nepal and Japan [19, 23, 28]. The obstructing membrane may be thick or thin, with or without a perforating ostium and is present at variable location in the intrahepatic portion of the IVC – above, at or below the level of the hepatic veins. There is invariably a concomitant narrowing of the IVC that resembles coarctation of the aorta.

This idiopathic membranous stenosis was originally postulated to be of congenital origin. However, the current view is that it is an acquired lesion and results from IVC thrombosis and subsequent scar formation. This hypothesis is supported by its late presentation, lack of anatomic or topographic consistency and clinical studies depicting evolution of IVC thrombosis into completely occluding membrane [12, 74]. Many patients with membranous stenosis have evidence of HBV infection (positivity for HbsAg) [74].

**IVC Thrombosis.** This invariably involves the intrahepatic portion of the IVC. Some of the probable factors for this phenomenon are intimal damage to the IVC by respiratory movements of the diaphragm and eddying of currents consequent to hepatic vein flow joining at right angles to the IVC flow.

**Oral Contraceptives and Pregnancy**

Pregnancy resembles a prothrombotic state due to increase in various clotting factors. The studies from India report pregnancy to be associated with 15–20% of patients of HVOO [19, 23]. In one of these studies, more than half of HVOO-related deaths occurred in pregnant patients [19]. Nearly 20% of cases occur in patients who have been on oral contraceptives [29–31].

**Uncommon Causes**

These include infections (liver abscess and hydatid cyst disease) and tumors (hepatocellular carcinoma, liver secondaries, hypernephroma, Wilm's tumor). These lesions cause outflow obstruction by compression and/or lead to the thrombosis of vascular structures [32]. In up to 20% of the patients, no cause of HVOO is detected. This percentage will decrease as more conditions such as factor V Leiden are increasingly diagnosed [33].

**Pathophysiology**

For HVOO to clinically manifest, a minimum of two or more hepatic veins should be blocked. Patients in whom only one hepatic vein is blocked may remain symptom-free [34]. Following hepatic vein outflow obstruction, the sinusoidal pressure rises and the sinusoids become dilated and congested. This causes necrosis of the centrilobular hepatocytes. In the early phase, HVOO clinically manifests as painful liver enlargement and ascites.

As the disease progresses, intra- and extrahepatic collaterals develop to decompress. This is more likely for the IVC obstruction than for hepatic vein obstruction.

The regions of the liver drained by the blocked hepatic veins atrophy while a compensatory hypertrophy takes place in regions with a patent venous outflow. The caudate lobe has multiple small hepatic veins that drain directly into the IVC. Thus, it usually escapes damage and undergoes hypertrophy to contribute towards hepatic function. This usually is a conspicuous feature on imaging studies in patients with BCS. The hypertrophied caudate lobe itself may cause pressure on the infrahepatic IVC and further aggravate ascites. The hypertrophied caudate lobe and high pressure in the infrahepatic IVC may be important considerations for the choice of shunt surgery for hepatic decompression for HVOO. However, in cases of isolated IVC obstruction, which is seldom complete, such caudate hypertrophy is not a constant feature.
Eventually the hepatocyte necrosis leads to cirrhosis. The natural history of an unrelieved hepatic congestion due to HVOO is therefore liver failure and death.

**Clinical Presentation**

The clinical differences between hepatic vein and IVC obstruction are presented in table 1.

Currently there is no universally accepted clinical staging system for HVOO. The system proposed by Bismuth and Sherlock [2] is commonly used. A patient with HVOO may present in one of the following four clinical stages [2, 19, 45].

**Fulminant**
This form is rare and has a high fatality. The patients present with fulminant hepatic failure with severe pain, tender hepatomegaly, ascites and encephalopathy within 8 weeks of jaundice. The fulminant form may complicate early puerparium [19].

**Acute**
The absence of encephalopathy distinguishes acute from fulminant form. The history is usually of short duration ranging between several days to 6 months. The patients have intractable ascites with tender hepatomegaly and splenomegaly as the collaterals have not yet developed to decompress the system. There is ongoing hepatocellular necrosis as evidenced by raised transaminases.

**Subacute**
These patients have a history usually ranging between 6 months to years. The hallmark is the development of collateral circulation that decompresses the hepatic sinu-

<table>
<thead>
<tr>
<th>Geography</th>
<th>IVC obstruction</th>
<th>Hepatic vein obstruction</th>
</tr>
</thead>
<tbody>
<tr>
<td>East</td>
<td>Membranous obstruction</td>
<td>West</td>
</tr>
<tr>
<td>Etiology</td>
<td>Long: months to years</td>
<td>Short: weeks</td>
</tr>
<tr>
<td>History</td>
<td>Male</td>
<td>Absent</td>
</tr>
<tr>
<td>Sex</td>
<td>Gross</td>
<td>Massive</td>
</tr>
<tr>
<td>Ankle edema</td>
<td>Moderate</td>
<td>Absent</td>
</tr>
<tr>
<td>Ascites</td>
<td>Anterior abdominal wall +</td>
<td>Tend, massive</td>
</tr>
<tr>
<td>Collaterals</td>
<td>Moderate/cirrhosis</td>
<td></td>
</tr>
<tr>
<td>Hepatomegaly</td>
<td>Good</td>
<td>Worse than IVC obstruction</td>
</tr>
<tr>
<td>Prognosis</td>
<td>Good</td>
<td>Worse than IVC obstruction</td>
</tr>
</tbody>
</table>

Investigations

The objectives of investigations in a patient suspected to have HVOO are confirmation of diagnosis, assessment of liver injury and delineation of the anatomy and the pressure profile of the block for treatment planning [14].

**Diagnosis of HVOO**
Abdominal Ultrasound with Doppler
This is the investigation of choice for initial detection and has a sensitivity and specificity of 75% or more [35–37]. The characteristic findings include obstruction of major hepatic veins at the juxta caval portion, hepatic veins devoid of signal, and/or segmental obstruction of the IVC. The commonly observed findings on Doppler ultrasonography include no flow or retrograde flow in the hepatic veins, presence of intrahepatic collaterals and reversed flow in the IVC.

CT Scan
The findings in the acute phase include increased liver volume, abundant free fluid and diffuse parenchymal hypodensity in unenhanced scan with strong post-contrast enhancement. The blocked hepatic veins are always demonstrable [38, 39]. The characteristic findings in chron-
ic cases include hypertrophy of the caudate lobe and left lobe, right lobe hypotrophy and hypoattenuation in the peripheral portion and bright inhomogeneous contrast enhancement in the hypertrophied areas. A segmental IVC obstruction is seen in 80% of the cases but membranous obstruction is seen only in 20% cases [40].

MR Imaging
The etiology of IVC obstruction is more likely to be diagnosed with MRI. The IVC may be obstructed due to caudate lobe hypertrophy, membrane or thrombus [41]. Other findings include hepatosplenomegaly, an enlarged caudate lobe and absent or attenuated hepatic veins with intraluminal thrombus. MR angiography may be indicated when the body habitus limits ultrasonography examination [42]. It may also be used to demonstrate the flow and anatomy of the portal vein and the IVC for the planning of PSS [43].

Conventional Angiography
This remains the gold standard for evaluating the pressure profile and the anatomy of the block for the planning of PSS.

IVC Venography. This investigation is performed to assess the patency of the IVC. The infrahepatic IVC is usually accessed through the transfemoral route. In patients with complete IVC obstruction, the length of obstruction may be assessed with a second catheter at the level of right atrium. During this study, pressure gradient above and below the block may also be recorded. A pressure gradient of 15 mm Hg or more is a prerequisite for mesoatrial shunt function [4].

Hepatovenography. This involves retrograde cannulation of the hepatic veins. The block may be classified as thrombotic (diffuse segments of the hepatic vein) or short segment stenosis which is usually a membrane or web located in the terminal part of hepatic vein [44]. This investigation also permits pressure measurements. A pressure gradient of 10 mm Hg or more between wedged hepatic vein pressure and the IVC is important for side-to-side portacaval shunt to function [45].

Assessment of Severity of Liver Damage
Liver Biopsy
The features diagnostic of the HVOO are severe and uniform centrilobular congestion and hepatocyte necrosis along with extravasation of blood in the space of Disse [46]. The biopsy should be bilobar as all three hepatic veins may not be equally affected by the thrombotic process. It is generally reserved for patients with subacute presentation. The findings of fibrosis or cirrhosis may provide impetus for transplant evaluation. In contrast, a patient with ongoing hepatocellular necrosis is a candidate for surgical shunt.

However, the pathologic interpretation of liver biopsy lacks standardization. Hepatic function is reported to recover after shunting in some patients where a preoperative liver biopsy demonstrated liver fibrosis [47]. This is corroborated by recent studies which show that liver fibrosis in early cases may be a reversible after the relief of biliary obstruction [48]. Liver biopsy is regarded by many as not an essential part of the diagnostic workup [1, 14, 20].

Other Investigations
The serum aspartate and alanine aminotransferase levels are usually elevated. In a patient with established cirrhosis, serum albumin may be low and other components (prothrombin time and serum creatinine) of the Child-Pugh scoring system may also be deranged. Most patients with HVOO have a high gradient ascites according to serum-ascites albumin gradient concept [49].

Management of HVOO
The objective of treatment in HVOO is to decongest the liver by providing adequate outflow in patients with ongoing hepatocellular necrosis. The main treatment modalities are discussed in the following sections.

Nonsurgical Management
Thrombolytic Therapy
This form of treatment may be considered for acute form of HVOO when the angiographic examination shows a fresh thrombus [22, 45, 50, 51]. The commonly used thrombolytic drugs are urokinase or tissue plasminogen activator and treatment is more effective when the drug is infused locally in the thrombosed vein. The experience with thrombolytic therapy for HVOO is limited and this modality may currently be regarded as investigational.

Interventional Radiology
TIPS. This involves the creation of an intrahepatic shunt between the hepatic vein and an intrahepatic portal vein via the percutaneous route. The objective is to achieve hepatic decongestion with the portal vein serving as the outflow tract.

The main indications for TIPS include failure of medical therapy for a patient with acute HVOO, where a por-
tacaval shunt is technically not feasible due to an enlarged caudate lobe or portal vein – infrahepatic vena caval pressure gradient is <10 mm Hg and as a bridge to liver transplantation in a patient with poor hepatic function [45]. For the latter indication, TIPS stent should not extend into the suprahepatic vena cava or portal vein as this may increase technical difficulties during recipient hepatectomy for transplantation [52]. TIPS placement has a procedure-related mortality of 1–2% and complications of approximately 10% [52]. In cirrhotic patients with variceal hemorrhage, new or worsened portosystemic encephalopathy manifests in 13–44% patients usually within 1 month of the procedure. The most important determinants are advanced liver disease and shunt diameter of >10 mm. It usually responds to standard medical regimen, shunt revision (reducing the diameter of shunt) may be necessary in few patients. The other late complication of TIPS is shunt dysfunction (defined as portal pressure gradient ≥12 mm Hg together with decrease in the luminal shunt diameter due to pseudointimal hyperplasia) which occurs in 18–78% patients [53]. The probability of shunt dysfunction increases with time with a majority of reinterventions being required by the end of 1 year. The results from recent series of TIPS for HVOO are presented in table 2.

### Table 2. Outcome of TIPS for HVOO

<table>
<thead>
<tr>
<th>Study (year)</th>
<th>n</th>
<th>Major complications</th>
<th>Stent dysfunction</th>
<th>Survival</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>Perello [66] (2002)</td>
<td>13</td>
<td>?</td>
<td>61% at a FU of 4–30 months</td>
<td>At a mean FU of 38 ± 26 months, 11 patients are alive and ascites-free</td>
<td>1 patient each died, has PSS or OLT</td>
</tr>
<tr>
<td>Mancuso [67] (2003)</td>
<td>15</td>
<td>Portal vein rupture 1, intrahepatic hemorrhage followed by rupture 1</td>
<td>40%</td>
<td>At a mean FU of 24 (8–44) months, 1 patient died (metastatic disease)</td>
<td>50% mortality in the acute liver failure group No patient needed OLT</td>
</tr>
<tr>
<td>Rossle [8] (2004)</td>
<td>35</td>
<td>20% (arteriostent fistulae 9%)</td>
<td>1-year probability 47%</td>
<td>1- and 5-year survival 93 and 74% respectively</td>
<td>2 patients needed OLT Survival data is transplant-free survival</td>
</tr>
<tr>
<td>Hernandez-Guerra [69] (2004)</td>
<td>25</td>
<td>Mild encephalopathy in 3 patients</td>
<td>81% at 1 year in the bare stent group vs. 33% at 1 year in PTFE-covered stents</td>
<td>At a median FU of 20.4 (range 3.9–124.8) months no patient died</td>
<td>Compared TIPS dysfunction in bare and PTFE-covered stents; 2 patients had OLT in the bare stent group.</td>
</tr>
</tbody>
</table>

Hepatic Vein Angioplasty. Hepatic vein angioplasty may be indicated for HVOO due to short length obstruction (see above). The procedure involves transjugular balloon angioplasty; in patients with severe residual stenosis a self-expanding metallic stent may be inserted. Recurrent obstruction occurs in 30–40% patients, many of whom can be salvaged by a repeat angioplasty [7]. Suboptimal anticoagulation may be one of the risk factors for re-occlusion. TIPS or portosystemic shunt are the modalities used to salvage the failures of hepatic vein angioplasty.

**Balloon Angioplasty with or without Stenting for IVC Stenosis.** This involves accessing the IVC through trans-femoral route and dilatation of the stenotic lesion till the ‘waist’ disappears. For focal lesions of the IVC like the membrane or web, satisfactory long-term results have been reported. In one of the studies, 24 of the 30 patients were symptom-free at a follow-up ranging from 6 months to 4 years [6]. Angioplasty with IVC stenting is indicated for recurrent stenosis after angioplasty or coarctation of the IVC [54, 55].

### Surgical Management of HVOO

**Portosystemic Shunts for HVOO**

The objective of PSS is to relieve hepatic congestion by converting the portal vein to an outflow conduit. The indications include acute or subacute HVOO where the investigations (elevated transaminases or liver biopsy) reveal an ongoing hepatocellular necrosis or failure of medical management to control ascites or variceal hemorrhage. The commonly performed shunts are side-to-side portacaval shunt, mesocaval shunt and mesoatrial shunt.

**Side-to-Side Portacaval Shunt (SSPCS).** This shunt may be performed for HVOO due to isolated hepatic vein thrombosis. The data on SSPCS for hepatic vein obstruction is rather limited, an earlier review reported only two studies with more than 10 patients each [5, 56]. The res-
ervation against performing SSPCS in patients with hepatic vein thrombosis is the likely technical difficulty in approximating the portal vein to the infrahepatic vena cava in the presence of a hypertrophied caudate lobe [2]. Another argument is that since a portacaval shunt involves hepatic hilar dissection, it may increase the technical difficulties if a subsequent liver transplantation is needed because the progression of liver disease [57]. Despite these reservations, excellent long-term graft patency and symptom-free survival of 81–94% has been reported in series from dedicated centers. The studies are summarized in table 3.

**Mesocaval Shunt (MCS).** MCS is the preferred shunt in the setting of isolated hepatic vein thrombosis in many studies [2, 4, 7]. The advantages of MCS are its technical simplicity and avoidance of hilar dissection. The prerequisite for this shunt is a patent IVC and it provides an effective hepatic decongestion even in those patients where an enlarged caudate lobe presses upon the IVC [2].

The shunt can be an autologous internal jugular vein or prosthetic (Dacron or PTFE). The main disadvantage of the Dacron grafts is high incidence (up to 50% or more in some series) of postoperative thrombosis whereas the reported long-term patency of the internal jugular vein grafts exceeds 80% [2, 3, 6, 58, 59]. The reported 5-year survival after MCSs for HVOO ranges between 57 and 95%. The results of the studies with MCS for HVOO are summarized in table 4.

**Table 3. SSPCS for HVOO**

<table>
<thead>
<tr>
<th>Study (year)</th>
<th>n</th>
<th>Perioperative complications</th>
<th>Survival %</th>
<th>Follow-up years</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pisan-Ceretti [56] (1998)</td>
<td>16</td>
<td>Shunt thrombosis in 6% of patients</td>
<td>81</td>
<td>5.7 ± 5</td>
<td>–</td>
</tr>
</tbody>
</table>

**Table 4. Mesocaval shunt for HVOO**

<table>
<thead>
<tr>
<th>Study (year)</th>
<th>n</th>
<th>Type of graft</th>
<th>Early PO results</th>
<th>Late results</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bismuth [2] (1991)</td>
<td>18</td>
<td>Internal jugular vein</td>
<td>In hospital mortality 1 patient († Procedure not specified)</td>
<td>5-year survival 95%</td>
<td>Early graft thrombosis salvaged by surgery in 2 (11%) patients All shunts patent at FU</td>
</tr>
<tr>
<td>Hemming [3] (1996)</td>
<td>12</td>
<td>Dacron 7 Internal jugular vein 1 Side-to-side 1</td>
<td>4 of the 21 PSS patients died</td>
<td>5-year survival 57% reported for PSS patients</td>
<td>4 of the 7 Dacron grafts thrombosed. With autologus internal jugular vein stent patency reported to be 80% at 2 months to 12 ears</td>
</tr>
<tr>
<td>Fisher [7] (1999)</td>
<td>16</td>
<td>Dacron/IJV (number not specified)</td>
<td>30-day mortality 3 (16%)</td>
<td>At a median FU of 67 months, 62% patients are surviving</td>
<td>Shunt block in 6 of the 13 (46%) patients at 2–94 months of FU. Of the 6 patients with shunt thrombosis 4 had Dacron and 2 IJV graft</td>
</tr>
<tr>
<td>Slakey [4] (2001)</td>
<td>24</td>
<td>Dacron/PTFE</td>
<td>30-day mortality 3 (12.5%)</td>
<td>Actuarial 5-year survival 75%</td>
<td>Primary and secondary shunt patency 70 and 83% respectively</td>
</tr>
</tbody>
</table>
Current Role of PSS in the Management of HVOO

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Thorax through an opening in the diaphragm and anastomosed to the right atrium via a right anterolateral thoracotomy [60, 61]. This is a technically demanding operation and long length of the synthetic graft, its placement in a low flow system and compression between the rigid liver and the thoracic wall make it prone for thrombosis [5, 62–64]. The reported primary patency of MAS varies between 22 and 46% [4, 6]. Cavoatrial shunt incorporates a side-to-side portacaval shunt with an added 20-mm diameter PTFE ring-reinforced shunt graft anastomosed to the side of the IVC at the level of the portacaval shunt and then anastomosed to the side of the right atrium. The results from one specialized center indicate that all the 12 patients receiving this graft are alive and symptom-free at a mean follow-up of 9 years [5]. The outcome of MAS for HVOO is summarized in table 5.

Other Surgical Procedures for HVOO. These include both SSPCS and cavoatrial shunt for the combined hepatic vein and IVC block, peritoneovenous shunt for the control of ascites (this does not reverse the sinusoidal congestion and hence is not recommended) and surgical excision of the IVC obstruction [5, 65]. The experience with these procedures is limited (except for the peritoneovenous shunt) with only a few specialized centers reporting good long-term results.

Liver Transplantation for HVOO

The accepted indications for liver transplantation in HVOO include fulminant hepatic failure, ESLD, decompensation after shunt surgery and inborn errors of metabolism like antithrombin III and protein C deficiency. The failure of previous surgical shunting procedures accounts for 15–25% of the indications for liver transplantation in HVOO patients [1]. The recent series from specialized centers have reported good results of liver transplantation for HVOO with 5-year patient survival of up to 95%. The results are summarized in table 6. In a recently published cumulative experience of 248 patients

<table>
<thead>
<tr>
<th>Study (year)</th>
<th>n</th>
<th>Graft</th>
<th>Perioperative mortality, %</th>
<th>Late results</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kohli [6] (1993)</td>
<td>11</td>
<td>Dacron 8, Ringed PTFE 2, Internal jugular vein 1</td>
<td>18</td>
<td>9 (81%) patients alive after 2–9 years. All 7 Dacron grafts became stenosed after an average of 2 years and needed reintervention.</td>
</tr>
<tr>
<td>Emre [68] (2000)</td>
<td>13</td>
<td>Ringed PTFE</td>
<td>15</td>
<td>76% patient symptom-free at median 42 months after surgery</td>
</tr>
<tr>
<td>Slakey [4] (2001)</td>
<td>19</td>
<td>Dacron/PTFE</td>
<td>16</td>
<td>Actuarial 5-year survival 75%</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Study (year)</th>
<th>n</th>
<th>Patient survival, %</th>
<th>Graft survival</th>
</tr>
</thead>
<tbody>
<tr>
<td>Jamieson [70] (1991)</td>
<td>26</td>
<td>69 69 50</td>
<td>–</td>
</tr>
<tr>
<td>Shaked [71] (1992)</td>
<td>14</td>
<td>86 76 –</td>
<td>2 patients had primary non-function and 1 had chronic rejection</td>
</tr>
<tr>
<td>Ringe [72] (1994)</td>
<td>43</td>
<td>69 69 69</td>
<td>–</td>
</tr>
<tr>
<td>Slakey [4] (2001)</td>
<td>6</td>
<td>– – 83</td>
<td>All the surviving 5 patients have functioning grafts</td>
</tr>
<tr>
<td>Srinivasan [9] (2002)</td>
<td>19</td>
<td>95 95 95</td>
<td>63% at a median FU of 83 (1–119) months</td>
</tr>
</tbody>
</table>
over a decade from 1988 to 1999 from 51 European centers, an actuarial survival of 71% has been reported at 5 years [73]. In this study, pretransplant renal failure and shunt were predictive of mortality.

### Follow-Up Management and Long-Term Results

Lifelong anticoagulation is recommended to maintain long-term shunt patency or after liver transplantation (except for inborn errors of metabolism) in patients with known coagulopathy [2, 5, 9]. The duplex Doppler ultrasound forms the investigation of choice for routine follow-up [4, 5]. In symptomatic patients with shunt occlusion, the different methods of salvage include clot lysis, percutaneous management including angioplasty and TIPS and revision surgery [2, 4–6]. The long-term outcome also depends on the management and natural history of primary blood disorders [9].

The long-term complications of PSS such as portosystemic encephalopathy, recurrent variceal hemorrhage or ascites for HVOO are rare (as compared to those for cirrhotic portal hypertension) and occur in the setting of shunt block or progression of liver disease. Excellent medium- and long-term results have been reported from specialized centers. In patients with functioning shunts with clinical disappearance of hepatosplenomegaly and ascites a normalization of liver function tests takes place usually within the first year following PSS [2, 5]. In the long term, in patients with patent shunts, hepatocellular function is well maintained and portosystemic encephalopathy has not been reported because of a normal functioning liver [2, 5, 75]. The patients who develop PSS dysfunction, not amenable to interventional radiological management or revision surgery and develop refractory ascites or progressive hepatocellular dysfunction may be considered for liver transplantation. The important causes of late deaths following PSS for HVOO include recurrence or progression of primary disease causing HVOO, shunt block and/or progression of liver dysfunction to ESLD [4].

### Discussion

HVOO is a rare disorder. The available literature on management has limitations of being retrospective, encompassing long time periods and small numbers [2, 4, 5]. Portosystemic shunts have been the mainstay of treatment with reported 5-year survival of patients ranging between 57 and 95% [2–5]. The limitations of PSS include declining expertise and postoperative mortality which ranges between 10 and 20% [3, 4, 7]. With advances in interventional radiology and liver transplantation and consequent availability of more treatment options, the role of PSS for the management of HVOO needs to be redefined.

Liver transplantation with reported disease-specific 5-year survival of up to 95% is currently the accepted treatment for the fulminant form of HVOO and for those patients with the chronic form with ESLD, decompensation after shunt surgery or unshuntable anatomy [9].

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Table 7. Treatment modalities for HVOO

<table>
<thead>
<tr>
<th>Presentation [2, 19, 45]</th>
<th>Level and type of obstruction</th>
<th>Preferred treatment modality</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fulminant</td>
<td>Hepatic vein or IVC thrombosis</td>
<td>Liver transplantation</td>
</tr>
<tr>
<td>Acute</td>
<td>Hepatic vein thrombosis</td>
<td>Thrombolytic therapy if investigations reveal a fresh thrombus</td>
</tr>
<tr>
<td></td>
<td>IVC thrombosis or long segment blocks</td>
<td>TIPS</td>
</tr>
<tr>
<td></td>
<td>Hepatic vein &amp; IVC thrombosis</td>
<td>TIPS</td>
</tr>
<tr>
<td>Subacute</td>
<td>Hepatic vein web or MOVC</td>
<td>Balloon angioplasty (± stenting)</td>
</tr>
<tr>
<td></td>
<td>Hepatic vein thrombosis</td>
<td>PSS (SSPCS/MCS)</td>
</tr>
<tr>
<td></td>
<td>Hepatic vein and IVC block</td>
<td>IVC stenting and SSPCS or MAS /CAS</td>
</tr>
</tbody>
</table>

End-stage liver disease: Liver transplantation

IVC = Inferior vena cava; MOVC = membranous obstruction of vena cava; TIPS = transjugular intrahepatic portosystemic shunt; SSPCS = side-to-side portosystemic shunt; MCS = mesocaval shunt; MAS = mesoatrial shunt; PSS = portosystemic shunt surgery; CAS = cavoatrial shunt.
TIPS is proposed as an alternative to surgical shunts. The currently acceptable indications for TIPS are: as bridge for liver transplantation for ESLD and liver decompression in sick patients of HVOO due to isolated hepatic vein thrombosis with acute presentation. The main limiting factor for this modality are procedure-related complications of approximately 10% and stent dysfunction of more than 50% of at 1 year [52]. The role of TIPS in patients with subacute presentation with ongoing hepatocellular injury remains to be clarified.

PSS is currently indicated for HVOO patients with subacute presentation and medically intractable ascites or refractory variceal hemorrhage but with preserved liver function. A recently proposed prognostic model has stratified patients into three groups (good, intermediate and poor prognosis) based on encephalopathy, ascites, prothrombin time and serum bilirubin. In this study, PSS was suggestive of improved survival only in patients with intermediate prognosis [22].

A 5-year survival of more than 90% has been reported after shunt surgery for HVOO in specialized centers [2, 5]. These results are superior to the currently available comparable figures for TIPS [8].

The aim of surgery is to have the portal vein serve as the outflow and thereby decompress liver. Two different shunts – SSPCS and MCS – have commonly been performed. The main reservation against performing SSPCS in the setting of HVOO is the technical difficulty likely to be caused by an enlarged caudate lobe. However, in the absence of any significant study that directly compares the two shunts and similar reported post-shunt 5-year survival, it is not feasible to make recommendation for any particular shunt and the choice is best governed by available local expertise [2–5]. One factor that favors MCS is the lack of hilar dissection during the performance of this shunt. MAS (or its variant cavoatrial shunt) is indicated for inferior cava blocks not amenable to interventional radiological management. The available treatment options and the current recommendations are summarized in table 7.

In conclusion, PSS, TIPS or liver transplantation are not competing but complimentary modalities for the treatment of HVOO. The choice is best determined by presentation, liver function tests, anatomy of block and the available local expertise.

References


Errata

In the article by Amit Nair, Dinker R. Pai and S. Jagdish entitled 'Predicting Anastomotic Disruption after Emergent Small Bowel Surgery' (Dig Surg 2006;23:38–43), in table 6 on p. 41, the correct range for the 95% CI for serum albumin should be 1.69, 16.67 (and not 0.06, 0.59 as printed) and for serum sodium 1.03, 1.21 (and not 0.82, 0.97 as printed).

The authors apologize for the error and any inconvenience this may have caused.

In the article by P.M. Brennan, T. Stefaniak, K.R. Palmer and R.W. Parks entitled 'Endoscopic Transpapillary Stenting of Pancreatic Duct Disruption' (Dig Surg 2006;23:250–254), the affiliation of Dr. T. Stefaniak has erroneously been stated as 'Department of Gastroenterological Surgery, City Hospital, Gdynia, Poland'. The correct affiliation however is 'Department of General, Endocrine and Transplant Surgery, Medical University of Gdansk, Poland'.


Current Role of PSS in the Management of HVOO


