Surgical Management of Noncirrhotic Portal Hypertension

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Noncirrhotic portal hypertension (NCPH) is a heterogeneous group of vascular diseases that lead to portal hypertension (PH) with normal or mildly elevated hepatic venous pressure gradient and preserved liver synthetic function. NCPH causes can be classified according to the site of portal flow resistance as prehepatic, hepatic, and posthepatic, with the hepatic causes further subclassified as presinusoidal, sinusoidal, and postsinusoidal. The main diseases producing NCPH are extrahepatic portal vein obstruction (EPVO), noncirrhotic portal fibrosis (idiopathic PH), hepatic vein outflow tract obstruction (Budd-Chiari syndrome), schistosomiasis, and congenital hepatic fibrosis. Manifestations of NCPH include upper gastrointestinal bleeding, ascites, portosystemic encephalopathy, hypersplenism, and portal biliopathy.¹⁻³

Timing and Indications of Surgery for NCPH

The main therapeutic goal in NCPH is to decrease portal pressure and thus prevent and treat PH complications. Surgery in NCPH aims to bypass the portal resistance site by creating portoportal or, more commonly, portosystemic shunts (PSS); when not feasible, direct treatment of complications of NCPH (eg, ligation of varices) may be undertaken. Surgery is generally reserved for patients with PH complications who do not respond to pharmacologic and endoscopic treatments but can also be indicated in patients with failure to thrive, limited access to healthcare, or desiring a definitive one-time treatment. Surgical PSS can help to treat portal biliopathy resistant to endoscopic treatment, but persistent biliary obstruction may need a biliary bypass. Some patients with symptomatic hypersplenism (such as repeated infections or bleeding, physical discomfort from

massive splenomegaly) may require simultaneous PSS and splenectomy.¹⁻³

Types of Surgical Procedures and Indications

Surgical procedures for NCPH can be divided into techniques that create a portoportal or a PSS and techniques that involve esophagogastric devascularization (direct ligation of varices or esophageal transection), with or without an associated splenectomy (Table 1). The most widely known of the esophagogastric devascularization techniques is the Sugiura procedure, which comprises splenectomy, devascularization of the superior two-thirds of the greater and lesser gastric curvature and the distal esophagus through the diaphragmatic hiatus (originally described with an abdominal and thoracic approach), and esophageal transection 4-6 cm above the gastroesophageal junction followed by end-to-end anastomosis (nowadays performed most commonly using a stapling device) (Fig. 1). Other techniques include the modified Hassab procedure (splenectomy with esophagogastric devascularization of the distal esophageal 7 cm and the proximal stomach with vagotomy and pyloroplasty), the Johnston procedure (only stapled esophageal transection and reanastomosis), and the Mathur procedure (transabdominal esophagogastric devascularization with gastroesophageal stapling and Nissen fundoplication with or without splenectomy).

PSSs can be classified as nonselective (decompress the entire portal system by diverting all portal flow), selective (decompress only the variceal-bearing compartment of the portal venous system and preserve some portal flow to the liver), and partial (incompletely decompress the portal system and maintain some portal flow).^{4,5} Nonselective PSS include end-to-side and side-to-side portocaval shunts (Fig.

Abbreviations: EPVO, extrahepatic portal vein obstruction; NCPH, noncirrhotic portal hypertension; PH, portal hypertension; PSS, portosystemic shunt.

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TABLE 1 Technical details of surgical procedures used in NCPH.

	Procedure	Indication
Esophagogastric devascularization techniques		Esophageal varices
Sugiura procedure	Distal esophagus & proximal gastric devascularization + Esophageal transection & reanastomosis + Splenectomy	
Hassab procedure	Distal esophagus & proximal gastric devascularization + Vagotomy & pyloroplasty + Splenectomy	
Johnston procedure	Esophageal transection & reanastomosis	
Mathur procedure	Distal esophagus & proximal gastric devascularization + Esophageal transection & reanastomosis + Nissen fundoplication +/- Splenectomy	
Portoportals shunts		Extrahepatic portal vein obstruction
Rex shunt	Connects superior mesenteric vein or varix to the left portal system at the recessus of Rex	
Portosystemic shunts		Esophageal varices Portal gastropathy Portal biliopathy
Nonselective (complete portal system decompression)		
Portocaval	End-to-side or side-to-side anastomosis	
Mesocaval	Side-to-side or with an interposition graft	
Mesoatrial	Requires interposition PTFE graft	
Proximal splenorenal	End-to-side anastomosis of proximal splenic vein to left renal vein, requires splenectomy	
Selective (variceal-bearing compartment decompression)		
Warren's distal splenorenal	End-to-side anastomosis of distal splenic vein to left renal vein, includes ligation of the coronary and pancreatic veins	
Inokuchi's left gastric venous caval	End to side anastomosis of left gastric vein vena cava, may require an autologous graft	
Splenocaval	End-to-side anastomosis of distal splenic vein to cava, may require an interposition PTFE graft	
Partial (incomplete portal system decompression)		
Sarfeh's portocaval	Uses 8 mm interposition PTFE graft

Figure 1 Sugiura procedure (from: Wright AS, Rikkers LF. Current management of portal hypertension. *J Gastrointest Surg* 2005;9:992-1005. They modified it from Sugiura M, Futagawa S. Further evaluation of the Sugiura procedure in the treatment of esophageal varices. *Arch Surg* 1977;112:1317.)

2A,B), mesocaval shunts (side-to-side or with an interposition graft) (Fig. 2C), and the proximal splenorenal shunt. Selective PSS include the distal splenorenal shunt (Warren shunt) (Fig. 2D), Inokuchi's left gastric venous caval shunt, and splenocaval shunts. Partial PSS includes the Sarfeh's small diameter interposition (8 mm PTFE graft) portocaval

shunt. The only portoportals shunt is the mesenterico-left portal vein bypass (Rex shunt), which restores the splanchnic flow to the liver, and thus its physiology and function, through the connection between the superior mesenteric vein and the portal system at the level of the recessus of Rex (Fig. 3).⁶

Selection of the surgical procedure depends on hepatic functional reserve and patency of the splenoportal venous axis, but also on patient's transplant candidacy and the available surgical expertise. Decompressive shunts are the best option to prevent rebleeding from esophageal and gastric varices and they can also obtain emergent bleeding control, although urgent shunting is rarely indicated. Devascularization procedures are indicated in patients with esophageal varices and unsuitable anatomy for a PSS. In schistosomiasis and idiopathic PH, distal splenorenal shunt is the technique of choice because of its effectiveness and lower incidence of postoperative hepatic encephalopathy compared to other PSS. Liver transplantation is the only cure for congenital hepatic fibrosis, but selective and nonselective PSS can often provide long-term benefit; partial liver resection may occasionally be needed in patients with heterogeneous involvement of the liver or recurrent cholangitis. In Budd-Chiari syndrome, appropriate initial anticoagulation can allow compensatory hypertrophy of uninvolved regions of the liver and thus long-term disease stability, but if anticoagulation fails, liver transplantation is the treatment

Figure 2 Non-selective portosystemic shunts: (A) end-to-side portocaval shunt; (B) side-to-side portocaval shunt; (C) end-to-end interposition mesocaval shunt; (D) distal splenorenal shunt. (from: Wright AS, Rikkers LF. Current management of portal hypertension. J Gastrointest Surg 2005;9:992-1005.)

Figure 3 Mesenterico-left portal vein bypass (Rex shunt) (from: Stringer MD. Improved body mass index after mesenterico-portal bypass. Pediatr Surg Int 2007;23:539-543.)

of choice. As a bridge to transplant, a side-to-side PSS (either mesocaval or portocaval) can be performed if the retrohepatic vena cava is patent, but cases where caudate hypertrophy results in caval occlusion require a long mesoatrial shunt. Mesocaval shunts are often preferred because of not needing hilar dissection and being technically simpler to construct, but late graft occlusion is common when a

prosthetic graft is employed because of the underlying hypercoagulability.⁵ Mesenterico-left portal vein bypass is, at present, the treatment of choice in EPVO because it restores the normal portal flow to the liver, although PSS (especially distal splenorenal shunt) can be used in cases of unfavorable anatomy.⁶

Outcomes After Surgery for NCPH

Shunt surgery effectively prevents rebleeding from esophageal varices and also improves portal gastropathy, portal biliopathy, and growth retardation.³ Overall prognosis and results of shunt surgery are much better in NCPH than in cirrhotic PH, with overall survival at 5 years around 80% and operative mortality <1%. Results depend on preoperative liver function status, need for an emergent procedure, postoperative patency of the shunt, and the specific disease (with EPVO showing the best long-term results). Main complications after shunt surgery are shunt occlusion, portosystemic encephalopathy, and rebleeding. Shunt occlusion depends on whether a graft (prosthetic or autologous) is used, with worse results using small diameter prosthetic grafts. Selective shunts like a distal splenorenal shunt result in a lower incidence of postoperative encephalopathy, but the selectivity of these shunts diminishes over time. Mesenterico-left portal vein shunt for EPVO has rebleeding

and portosystemic encephalopathy rates <5%, nearly absent mortality, and shunt patency between 75% and 100%.⁶ In schistosomiasis, distal splenorenal shunt is superior to devascularization procedures with lower rebleeding risk (5%-22%) and better survival (around 90%), but with a higher encephalopathy rate (10%-15%).⁷ Distal splenorenal shunt for idiopathic PH has low mortality rate (0.8%), low variceal rebleeding rate (8%), and low post-shunt encephalopathy rate (13%).³ In congenital hepatic fibrosis, distal splenorenal shunt shows absent mortality, minor morbidity, low shunt occlusion (<20%) and moderate rebleeding rates (up to 20%).⁸ The role of shunt surgery in Budd-Chiari syndrome is least well-established; shunts present a dysfunction rate as high as 32%, along with high morbidity and mortality rates, although survival rates after shunting range between 57% to 94%.⁹

Summary

Surgery in NCPH is reserved for patients with PH complications who fail medical and endoscopic treatments. Surgery

aims to disrupt esophagogastric collateral circulation through devascularization procedures or to decrease portal pressure by creating portoportal or PSS. Devascularization procedures are useful in patients with esophageal varices and unfavorable anatomy for a shunt. The only portoportal shunt is the mesenterico-left portal vein bypass (Rex shunt) and it restores the normal portal flow to the liver in EPVO. Decompressive PSS (including nonselective, selective, and partial) prevent rebleeding from esophageal and gastric varices but they may complicate with portosystemic encephalopathy, occlusion, and subsequent rebleeding. Results after shunt surgery depend on the specific disease (with EPVO showing the best long-term results), preoperative liver function status, and postoperative patency of the shunt.

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