

*Editorial***Surgery for Non-cirrhotic portal hypertension: Current status****Vivek Mangla¹, Sujoy Pal², Peush Sahni²**

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Non-cirrhotic portal hypertension (NCPH) is a term used to describe a heterogeneous group of diseases characterized by the presence of portal hypertension with preserved liver structure and function. It is a common cause of portal hypertension in children (up to 70%) and young adults in India and other developing countries. The morbidity is mainly related to variceal bleeding, hypersplenism, growth retardation and portal biliopathy. There are two distinct disease entities resulting in NCPH, namely, extrahepatic portal venous obstruction (EHPVO) and non-cirrhotic portal fibrosis (NCPF). This distinction is largely based on the presence of a portal cavernoma replacing the thrombosed portal vein in the former, and a patent dilated spleno-portal venous axis in the latter.

Endoscopic management (sclerotherapy or variceal ligation) combined with some form of pharmacotherapy is the treatment of choice for acute variceal haemorrhage. However, about 5% patients may need emergency surgery for control of acute variceal haemorrhage following failure of medical and endoscopic management.¹ Studies have shown that oesophagogastric devascularization, without oesophageal stapled transection, is a safe and effective procedure for control of variceal bleeding with similar results and lesser morbidity than devascularization with oesophageal transection.² Nevertheless, considering the somewhat higher variceal re-bleeding rates (>20%) in patients treated with oesophagogastric devascularization (according to Zhang)³, over a period of time with increasing experience, portosystemic shunt has become the procedure of choice in this setting.⁴

The main indication for surgery in patients with NCPH is prevention of variceal haemorrhage (i.e. secondary prophylaxis). The efficacy of endoscopic treatment for secondary prophylaxis has been reported by various series.^{5,6} Even though, in the long term, largely similar re-bleeding rates have been reported with endoscopic treatment and surgery (17% vs. 11%), endoscopic treatment requires stringent long-term follow-up. In addition, repeated sessions for obliterating varices can lead to oesophageal strictures (~15%), development of ectopic varices and congestive gastropathy. Also, during the time that varices are being eradicated (about 8-12 weeks), the risk for variceal haemorrhage remains. Endoscopic therapy, further more, does not reverse growth retardation, hypersplenism, portal biliopathy or recurrent pain due to infarcts in the enlarged spleen. Yet, these always

improve following shunt surgery (especially after splenectomy and proximal lienorenal shunt [LRS]).

Broadly, the surgical options for these patients can be divided into portosystemic shunts, oesophagogastric devascularization and the more recently described Rex shunt.

Portosystemic shunts are usually feasible in about 90% of patients. The options for portosystemic shunts include proximal LRS, side-to-side LRS, distal LRS and mesocaval shunt. The side-to-side LRS preserves the spleen. However, following this procedure, hypersplenism may take a long time to reverse or may not reverse at all. It is also not suitable for patients with massively enlarged spleens and in those experiencing recurrent pain. Distal LRS, though preferred in patients with cirrhosis, does not offer any advantage in EHPVO as encephalopathy does not occur even after non-selective portosystemic shunts in these patients.^{7,8} Removal of the spleen as part of a proximal LRS helps treat hypersplenism as well as recurrent pain. There is no major disadvantage from removing the spleen as overwhelming post splenectomy infections are rare in these patients.^{7,8} A mesocaval shunt usually requires the use of a vascular conduit (autologous internal jugular vein or a synthetic graft) and has higher rates of shunt thrombosis. Among 160 children who underwent proximal LRS, the overall mortality was 1.9% (elective: 1/140 [0.7%]; emergency 2/20 [10%]), re-bleeding rate was 11% and actuarial survival at 15 years was 95%.^{6,7} Long term studies (15-40 years) from western countries have repeatedly shown that shunt surgery with or without splenectomy (proximal LRS, side-to-side LRS, distal LRS or mesocaval) for EHPVO in children and adults, is associated with shunt patency rates between 90% and 95%, re-bleeding rates of 5%-10%, no encephalopathy and 15-year actual survival rates of 95%.^{5,9-11} Good long-term results in terms of low morbidity and mortality (as low as 1%) and low re-bleeding rates (as low as 4%) have been reported with portosystemic shunts even in very young children.⁷

Devascularization entails ligation of varices or veins communicating with these varices near the lower oesophagus and proximal stomach. Even though splenic preservation has been reported in devascularization, splenectomy facilitates the procedure, and reverses

hypersplenism without any major detrimental effects. Since there is no decompression of the portal venous system, portal pressures remain high with the attendant risk of worsening portal biliopathy, portal hypertensive gastropathy with an additional risk of development of ectopic varices. Devascularization is associated with re-bleeding rates of 11%-17% in patients with EHPVO.^{9,10}

The Rex (mesenterico-left portal vein) shunt is done using a conduit (autologous internal jugular vein or synthetic graft) to restore blood flow from the superior mesenteric vein to the intrahepatic left portal vein, bypassing the block in patients with EHPVO. It is more physiological with potential advantages in terms of improvement in the size of the left lobe of the liver, cognitive ability and reversal of hypersplenism.^{11,12} However, the extent to which these advantages are realized remains to be seen with one series showing that the procedure was feasible in only 60% of patients selected for surgery.¹³ The fact that most early series of Rex shunt included children, many of them in the post-living donor liver transplantation setting, who had developed portal vein thrombosis that was probably detected early, suggests that this may not be applicable to older patients with EHPVO. We have observed that in many patients with EHPVO the thrombotic obliteration extends into the left portal vein making a Rex shunt a non-viable option. Two head-to-head comparisons between Rex shunt and non-selective portosystemic shunts are available. These do not show a clear advantage of the Rex shunt over other porto systemic shunts in patients with NCPH.^{13,14}

A group of patients with EHPVO with certain high risk features with no previous variceal bleeding have also been treated with 'prophylactic' surgery (mostly portosystemic shunts) with good outcomes.¹⁵ The prophylaxis is for variceal bleeding and quite often some other indication for surgery is usually present in these patients including recurrent pain from an enlarged spleen, hypersplenism, portal biliopathy, portal hypertensive gastropathy or growth retardation. The long-term results are good with low re-bleeding rates, no overwhelming post-splenectomy infections or post-shunt encephalopathy.¹⁵ In view of these, surgery offers the potential for a one-time cure in these patients.

Surgery for EHPVO has evolved from largely emergency control of variceal bleeding using mainly

devascularization procedures, to elective shunt surgery being the preferred treatment for most patients. It is even beneficial in patients with EHPVO without history of previous variceal haemorrhage.¹⁶

The long-term results of shunt surgery for NCPF are good in terms of high patency and low re-bleeding rates (3%-19%), attributable to the good size of the splenic vein.^{16,17} However, a higher incidence of encephalopathy (2%-13%) has been reported following shunt surgery in these patients.^{16,17} Furthermore, though uncommon, long-term complications such as hepatopulmonary syndrome (postulated to result from pulmonary arteriovenous fistulae), glomerulopathy and myelopathy have been reported after shunt surgery in these patients.¹⁸ Though a higher rate of re-bleeding has been reported with devascularization compared to shunt surgery, it has certain advantages as encephalopathy, glomerulopathy and myelopathy do not occur in these patients.^{9,10} In view of these results, it is prudent that patients with NCPF are carefully evaluated for the indication of surgery. Patients with failed endotherapy who are at high risk of re-bleeding should undergo a portosystemic shunt such as a proximal LRS. However, those who have not had variceal bleeding usually have low grade varices and require surgery for hypersplenism or pain related to massive splenomegaly. These patients should be managed with splenectomy and devascularization.

A recent series has reported that the results of portosystemic shunt may not always be good and that shunt patency rates tend to decline with time.¹⁹ This experience is different from previously reported series and may be related to the relatively smaller number of patients studied. It is only in the early follow up period that shunt blocks can be ascribed to technical factors. Decline in shunt patency rates with time seems to suggest that non-technical reasons may be responsible for this. A study has suggested that nearly one-third of patients with EHPVO have some inherited or acquired pro-coagulant tendency.²⁰ Hence, it may be useful to study whether anticoagulation will help increase the long-term shunt patency rates. A better understanding of the etiology of EHPVO and NCPF is needed to help identify patients at increased risk of shunt block and selective use of anticoagulation may help increase shunt patency rates in

the long-term. In India, the expertise to provide effective endoscopic variceal obliteration is probably more widely available than for shunt surgery. Also, endoscopic variceal obliteration is an effective modality to treat acute variceal bleeding, manage patients with re-bleeding due to blocked shunts as well as to treat varices in patients who are not eligible for shunt surgery because of non-availability of a suitable vein.

Thus, in the present era of TIPS and liver transplantation, shunt surgery and devascularization will continue to hold an important position in the management algorithm of patients with NCPH.

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