Surgical treatment of Budd-Chiari syndrome

Budd-Chiari syndrome (BCS) is a relatively uncommon cause of portal hypertension, constituting 7% to 9.1% of cases in India.1,2 In Western countries, it is usually a sequel of obstruction in the main hepatic veins and a definite cause can be determined in most cases.1 However, in countries like India, Nepal, China and Japan, obstruction to the suprahepatic inferior vena cava (IVC) is more common and an underlying hypercoagulable state is identifiable in only a small percentage of patients.3-9

Hepatic venous outflow obstruction leads to centrilobular congestion and necrosis of the hepatic lobule, ascites and finally to cirrhosis. The prognosis of untreated cases remains poor as spontaneous resolution of obstruction is rare. The reported mortality with hepatic vein thrombosis is 22%-42% at 1 year and 51%-90% at 3 years.10 The corresponding figures for IVC obstruction are 30% at 1 year and 60% at 2 years. Treatment options for Budd-Chiari syndrome include medical, radiological and surgical techniques; however, treatment remains difficult and unsatisfactory.

Medical measures for control of ascites are meant for palliation only and probably do not alter the course of the disease. There are reports of successful thrombolytic therapy with streptokinase, urokinase and tissue plasminogen activator; the experience is very limited.11,12 Additionally, thrombolysis almost invariably fails in chronic cases.13 Anticoagulation without radiological or surgical intervention usually fails.

Radiological interventions include balloon angioplasty with or without vascular metallic stent placement for membranous obstruction or short-segment thrombotic obstruction of the IVC.13-16 Experience with angioplasty procedures for hepatic vein obstruction is very limited. Laser-assisted angioplasty has been used lately with good results.17 Radiological intervention procedures have emerged as preferred therapeutic modality for membranous obstruction or short-segment obstruction of the IVC at most centers.5

Surgical options for patients with BCS include minor palliative procedures like peritoneovenous shunts, various shunt procedures, procedures aimed at promoting collateral circulation, e.g., splenopneumoperexy, omentostomopexy, omentorenopexy, etc.; direct surgical procedures like transcardiac membranotomy, transscapular liver resection with hepatoatrial anastomosis, etc.; and liver transplantation. Experience with many of these surgical procedures has been limited and results reported by different groups of workers have been variable.

In this issue of the Journal, Shah et al16 describe their experience with surgical treatment of Budd-Chiari syndrome. Twenty-three of 61 of their patients were not offered any surgical procedure because of evidence of cirrhosis; another 22 patients were offered radiological intervention procedures, details of which have not been provided.

Evidence of cirrhosis or poor hepatic functional reserve precludes any shunt procedure because of high risk of postoperative complications and encephalopathy. Such patients are candidates for liver transplantation.13,16,19,20 However, availability of liver transplantation facilities is limited in developing countries like India.

A large number of shunt procedures have been described; this reflects the lack of consensus about the best surgical approach in such patients. Side-to-side portocaval shunt is considered the procedure of choice if IVC pressure is substantially lower than portal pressure.21,13,16,21,22 This procedure is associated with an operative mortality of as low as 5% and prolonged survival in up to 90% of patients (follow up of more than 8 years).13,21-24 However, the operative procedure can be demanding in BCS due to the enlarged caudate lobe. Measures suggested to overcome this include proper positioning of the patient, proper mobilization of the portal vein and IVC, and rarely partial resection of the caudate lobe.21 Another option is a two-step procedure — mesoatrial shunt to decongest the liver followed by side-to-side portocaval shunt after the enlarged caudate lobe decreases in size. However, such an approach has not found wide favor.

In the present study, side-to-side portocaval shunt was performed in 3 patients; these shunts remained patent till the last follow up. Such a shunt was precluded in 3 other patients because of enlarged caudate lobe (one patient), aberrant course of hepatic artery (one patient) and IVC stent (one patient). If the IVC is patent and a gradient of > 20 mmHg exists between the portal vein and IVC and a portocaval shunt cannot be performed due to any of the above mentioned reasons, mesocaval shunt with a graft is probably the best option.24-28

Mesocaval shunt is less demanding and does not interfere with subsequent liver transplant.16 Various types of grafts used for mesocaval shunt include the jugular vein, Dacron and PTFE with reinforced rings.28 In the present study, mesocaval shunt was performed in only one patient using PTFE graft; this remained patent during follow up. Two patients were subjected to porto-renal shunt and both died within two weeks of surgery. Experience with such a shunt is very limited. Other shunts involving the renal vein, e.g., splenoportal shunt, have been reported to give poor results.

Surgical therapeutic options for combined hepatic vein and IVC block are limited. Mesoatrial shunt and one-stage side-to-side portocaval shunt with cavo-triatrial shunt are the two procedures which are generally recommended.13,15,16,25,27,28 Both these require the use of syn-
thetic grafts, namely, ring-enforced PTFE grafts. These grafts increase the risk of shunt blockage. In the present study, mesoatrial shunt was performed in two patients; both got blocked or infected.

Transcaval liver resection with hepatointestinal anastomosis (Senning’s procedure) has been described for hepatic venous block or combined hepatic venous and IVC block. This procedure has the theoretical advantage of restoration of normal drainage and has been shown to give good results during long-term follow up (up to 11 years, mean 6). However, experience with this procedure is limited to a few centers.

In the present study, gastric devascularization was done in one patient. This was essentially a palliative procedure for bleeding gastric varices and does not alter the natural history of hepatic venous outflow tract obstruction.

Thus, most of the patients in the present study were treated by radiological intervention which remains the primary modality of treatment (if technically feasible) for membranous or short-segment obstruction of IVC and short-segment obstruction of hepatic veins.

The follow up period in the present study is short (mean 21.7 months). The degree of preservation of hepatic function in the long term despite a patent portosystemic shunt has been reported to be variable. Shunt procedures which are based on the presumption that the portal vein will function as outflow tract do not reverse hepatic congestion completely. It is well known that some patients continue to deteriorate despite patent portosystemic shunts and need liver transplantation ultimately.

Every patient with BCS needs to be investigated for underlying hypercoagulable state or other causes leading to hepatic outflow tract obstruction. The underlying cause must be treated appropriately. The present study is silent on this aspect. Long-term anticoagulation is recommended for any patient subjected to portosystemic shunt for BCS with underlying hypercoagulable state, as the risk of thrombotic complications is high in these patients.

Although the number of patients in the present study is relatively small, the overall results are encouraging. The results are likely to improve in future with increasing experience of the surgeon and better understanding of the subject.

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References

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