Management of the Budd-Chiari syndrome by balloon cavoplasty

BINAY K DE, PRANAB K BISWAS,* SAMBIT SEN, DEBASISH DAS, KALLOL K DE, UTPALENDU DAS,** SANJAY K MANDAL, DEBASISH MAJUMDAR

Departments of Medicine, *Cardiology and **Radiology,
Institute of Postgraduate Medical Education and Research, 244 Acharya JC Bose Road, Kolkata 700 020

Background: Obstruction of the suprahepatic inferior vena cava (IVC) by a membrane or stricture is the commonest cause of Budd-Chiari syndrome in the eastern hemisphere. We present our experience with the outcome of balloon cavoplasty in such cases. Methods: We followed up 40 consecutive cases of Budd-Chiari syndrome over seven years. Doppler study of hepatic venous outflow tract (in all cases), liver biopsy (30 cases) and necropsy (two cases) were performed. Balloon cavoplasty was done in selected cases. Results: Of 40 patients with BCS (mean age 35.2 [SD 8.7] years; 26 men) 5, 5 and 30 had fulminant, acute and chronic presentation, respectively. Inferior vena cavaography was performed in 32 cases, and showed membranous obstruction of the IVC in 12, segmental occlusion of the IVC in 11 cases, and block in both the IVC and the main hepatic veins in the rest. Successful balloon cavoplasty was done in 18 cases with obstruction of the IVC (membrane or stricture); 15 of them are well over a mean follow up of 56 (14.6) months. Three patients developed restenosis; two of them, treated with redilatation, are doing well, and one died of septicemia and hepatic failure following a surgical bypass. Pressure gradient between the IVC and right atrium decreased significantly after cavoplasty (15.4 [2.8] vs 6.6 [2.0] mmHg; p < 0.001). Conclusion: Balloon cavoplasty gave encouraging results in the management of Budd-Chiari syndrome due to membranous obstruction or stricture of the IVC. [Indian J Gastroenterol 2001;20:151-154]

Key words: Hepatic venous outflow obstruction, inferior vena cava, membranous obstruction, cavoplasty

There are distinct differences in the underlying conditions and in the anatomic pattern of Budd-Chiari syndrome (BCS), between the eastern and western parts of the world.1 Whereas hematologic disorders are the leading cause of BCS in Western countries, stricture of the inferior vena cava (IVC) and idiopathic membranous obstruction of the IVC (MOVOC)2 are the most common causes in the eastern hemisphere. The latter lesions have been described from Japan,3 South Africa4 and India,5 and include lesions varying from a thin web-like membrane to a thick fibrotic band, with or without associated thrombus, obstructing the IVC at or above the level of the hepatic venous ostia.

Various surgical procedures, including direct approach and bypass operations,6 have been attempted for this disease but with significant morbidity and mortality. Balloon cavoplasty has been recommended as the treatment of choice, with good results7-10 especially in patients with isolated IVC membrane or stricture leading to BCS. Our aim was to evaluate the efficacy of cavoplasty in BCS due to MOVOC and stricture of IVC.

Methods

Patients with clinical features suggestive of BCS, like ascites, hepatomegaly, tortuous veins on the anterior abdominal wall or the back, pedal edema, or jaundice, and those incidentally found to have hepatic venous outflow obstruction (HVOC; n = 2) during work-up for other conditions were evaluated. Patients with cardiac diseases like constrictive pericarditis and those with veno-occlusive disease were excluded.

Forty consecutive cases with BCS were included in our study over a 7-year period (1992-1998) from patients attending the Portal Hypertension Clinic and those admitted in the medicine indoor wards of our institute. After clinical examination, patients were investigated by hemogram, liver profile, coagulation profile (including prothrombin time in all patients, and protein C and protein S measurement in 6 patients), serum alpha fetoprotein, viral markers (HBsAg, anti-HCV), and upper gastrointestinal endoscopy. Ultrasonography with Doppler study (EUB-515A ultrasound scanner with 3.5 MHz electronic convex transducer; Hitachi, Tokyo, Japan) was done in all cases with emphasis on hepatic venous outflow tract. Computed tomographic scan of abdomen was performed in 10 cases, either because of a suggestion of malignancy (hepatocellular carcinoma - 6, cholangiocarcinoma - 1) or pancreatitis (1), or to rule out extrinsic compression of the IVC by an enlarged caudate lobe (2). Liver biopsy using a Tru-cut needle was done in 30 cases and at necropsy in two cases. The study protocol was approved by the Ethics Committee of our institute.

The procedure for hepatic vein and IVC catheter-
ization was explained to the patients and an informed consent obtained. After an overnight fast, IVC catheterization was done using the Seldinger technique through the right femoral vein, and the right antecubital vein if necessary, under local anesthesia. A 7F pigtail catheter (CUSA; CR Bard Ireland, Galway, Ireland) was introduced under fluoroscopic guidance (Polydiagnost C2; Philips, Best, the Netherlands) and inferior venacavography was done by digital subtraction angiography. Site and nature of the IVC block were recorded.

Cavoplasty was considered in patients who fulfilled all the following criteria: (i) either MOVC or IVC stricture, (ii) adequate hepatic reserve, and (iii) at least two patent main hepatic veins and patent portal vein. Patients were excluded in case of (i) refusal of consent, (ii) associated malignancy, (iii) blockage of more than one main hepatic veins, or (iv) extrinsic compression of the IVC. Using an end-hole 7F universal catheter, IVC pressure, and free and wedged hepatic venous pressures (whenever possible) were recorded. Then, a guide wire (~0.89 mm in diameter) was introduced through the catheter and maneuvered through the stenosed segment under fluoroscopic guidance. If the obstructed segment could not be negotiated, venography through the right antecubital vein was performed and the extent of obstruction was assessed. In cases with MOVC, the stiff end of the guide wire was used to puncture the membrane. In six cases in whom this failed, Brockenbrough’s trans-septal needle was used to puncture the obstructing membrane. After negotiating the obstructed segment, pressures in the IVC above the obstruction and in the right atrium were recorded using a multichannel pressure recorder (Howlett Packard universal quartz transducers HP 1290C; Palo Alto, California, USA). Following this, an Inoue mitral valvuloplasty balloon (22 mm, single balloon; Toray Medical, Tokyo, Japan) was negotiated across the obstructed segment, and inflated and deflated (2-4 times with incremental pressures to achieve graded dilatations) till disappearance of the waist. The balloon was then removed, and IVC venography and measurement of IVC and right atrium pressures were repeated. Technical success was defined as (i) decrease in pressure gradient, and (ii) >50% increase in vessel diameter on angiography.11 The patients received anticoagulants for a period of at least 3 months; initially, intravenous heparin was given for 3-4 days to achieve an APTT 2-2.5 times the normal value, followed by oral warfarin to maintain a prothrombin time of 2-2.5 times normal.

Patients were followed up clinically. Liver function profile, upper gastrointestinal endoscopy and Doppler study were repeated after 6 months. Restenosis was suspected if clinical features of BCS reappeared; it was then confirmed using ultrasound Doppler and angiography. Redilatation was attempted using the same method as above.

Data are expressed as mean (SD). Student’s t test was used as applicable; p<0.05 was taken as significant.

Results
The mean age of the 40 patients (26 men) was 35.2 (8.7) years (range 12-60). Five (12.5%) patients each presented as fulminant and acute, and 30 (75%) as chronic, BCS. Clinical characteristics of our patients are shown in the Table. Liver biochemistry profile showed bilirubin 2.6 (2.2) mg/dL, AST 110 (46.5) IU/L and ALT 52.9 (6.8) IU/L; prothrombin time ranged from 13 to 20 seconds (control 12-13). One patient had protein S deficiency (protein S antigen level 46.8% [normal 73-151]). Serum albumin was low (<3.5 g/dL) in 24 cases; 13, 24 and 3 patients belonged to Child’s class A, B and C, respectively. HBsAg was positive in 4 patients, all of whom had hepatocellular carcinoma. Esophageal varices were present in 34 cases (grade I, II, III and IV in 13, 15, 5 and 1 patients, respectively).

Ultrasoundography showed features consistent with BCS in all cases. In addition, space-occupying lesions in the liver were detected in 7 patients (suggestive of hepatocellular carcinoma in 6 and hydatid cyst in 1), extrahepatic biliary obstruction in one (diagnosed as cholangiocarcinoma on histology), chronic pancreatitis in one, and portal vein thrombosis in 3 patients. Histology was available in 32 cases (biopsy - 30, necropsy - 2); it revealed cirrhosis in 10, centrilobular congestion and necrosis in 9, mixed picture in 6 and malignancy in 7 patients. Eight of these (hepatocellular carcinoma - 6, cholangiocarcinoma - 1, hydatid cyst - 1) were not subjected to angiography. IVC angiography done in the remaining 32 patients revealed MOVC in 12 patients and partial segmental occlusion of the IVC in 11; 9 cases had IVC and hepatic vein block — stenosis of IVC and hepatic veins in 7, thrombi at multiple sites (confirmed by ultrasound Doppler study) in 2 cases.

Ballooon cavoplasty was attempted in 23 patients (MOVCl2, IVC stricture I) and could be successfully performed in 18 cases (MOVCl, IVC stricture 7),

| Table: Clinical characteristics of patients with Budd-Chiari syndrome (n=40) |
|---------------------------------|-----------------|-----------------|
| Clinical feature               | No. of cases    | Prevalence (%)  |
| Hepatomegaly                   | 38              | 95              |
| Anterior abdominal wall veins  | 38              | 95              |
| Ascles                         | 30              | 75              |
| Pain in abdomen                | 28              | 70              |
| Back vein                      | 34              | 85              |
| Splenomegaly                   | 26              | 65              |
| Ankle edema                    | 28              | 70              |
| Jaundice                       | 15              | 38              |
| Gastrointestinal bleeding      | 6               | 15              |

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with an overall success rate of 78.3%. In these latter cases, repeat inferior venacavography showed disappearance of the waist produced by the obstructing segment (Fig). Cavoplasty was not successful in one patient with MOVC (thick membrane that could not be punctured) and 4 patients with IVC stricture (segments >4 cm in length, which could not be successfully negotiated).

Inferior vena cava pressure (below obstruction) before and after cavoplasty was 19.9 (3.0) and 11.6 (2.6) mmHg, respectively (p<0.001). The mean pressure gradient between the right atrium and the IVC was 15.4 (2.8) mmHg before the procedure and decreased to 6.6 (2.0) mmHg after the procedure (p<0.001).

The patients were followed up for 56 (14.6) months. Among patients with successful cavoplasty, anterior abdominal and back veins persisted in 2 of 18 cases each, ascites in 1 of 18 cases, pain in abdomen in 2 of 16 cases and jaundice in 1 of 10 cases. No patient had persistent ankle edema or further episodes of variceal bleed. Re-evaluation after 6 months revealed an overall improvement of Child class A: B: C = 14:4:0, from 8:10:0 initially) and a reduction in variceal size (variceal grade none: I:II:III:IV = 10:5:2:1:0, from 2:6:8:2:0 initially). Fifteen patients were well with no further need for dilatation. Three patients developed restenosis (after 9, 16 and 11 months). Two of them are well after redilatation. One patient was treated with surgically bypass (after failure of repeat cavoplasty, due to the tight nature of the stenosis), but died of sepsis and hepatic failure after surgery.

Among the patients in whom cavoplasty was not performed, all 7 cases with malignancy died, whereas the patient with hydatid cyst underwent surgery and is doing well. Of the remaining 14 cases, only 5 attended the follow-up clinic; 2 of them have died (variceal bleed after 3 months in one, bacterial peritonitis after 7 months in the other). Another patient is in a terminal condition, whereas the other 2 remain in an unchanged condition (8 and 14 months later).

Discussion
IVC obstruction can occur due to several underlying defects, such as congenital malformations, neonatal obliterator changes in the veins, acquired thrombophlebitis, and a combination of these factors. In a study of 44 cases with IVC obstruction, the underlying pathophysiology defect could not be determined in any.13 Our angiographic findings reveal that the obstruction may be short in length and truly membranous or may be a segmental occlusion extending for several centimeters. The site of the obstruction was nearly always at the level of the diaphragm. Obstruction of the IVC differs from that of the hepatic veins in several respects, viz., (a) the clinical manifestations develop more insidiously, (b) prominent subcutaneous truncal collaterals are almost always present, (c) chronic infection with hepatitis B virus is common. In our study only 4 (10%) patients were positive for HBsAg.

The outcome of IVC block is somewhat poor because of the high incidence of cirrhosis and hepatocellular carcinoma (48.5% in one study). Various surgical procedures including direct approach and bypass operations have been attempted in this disease and portosystemic shunt has been considered as a life-saving procedure. However, its cost, morbidity, chance of thrombosis of shunt and high operative mortality make it unsatisfactory. With patent shunt, 5-year actuarial survival rate is 87%; however the survival rate is as low as 38% in those with shunt thrombosis. Early portal decompression is mandatory, with low rate of success and with mortality of 40% in some series.

A recent report recommended that the use of surgical shunt procedures should be restricted to the management of refractory ascites or variceal bleeding in BCS.7 Balloon dilatation is particularly valuable if only the IVC is obstructed and multiple dilatations are usually necessary. Because of the limited role of conservative treatment for IVC obstruction due to membrane or stricture, we undertook balloon cavoplasty in all our cases fulfilling the criteria mentioned. Cavoplasty resulted in significant venous decompression as evidenced by reduction in the pressure gradient. Thus, the progressive damage due to back pressure effect of hepatic venous outflow tract obstruction is probably halted. Whether the associated hepatic parenchymal damage is reversed needs further evaluation.

We conclude that balloon cavoplasty is an effective treatment modality for patients of BCS due to MOVC or stricture of the IVC in the absence of hepatic venous obstruction.
References


Correspondence: Dr B K De, 63/4A/1A, Dr SC Banerjee Road, Kolkata 700 010. Fax: (33) 476 1799. E-mail: biney.kde@apexmail.com

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