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Original Article

Budd-Chiari syndrome managed with percutaneous recanalization: Long-term outcome and comparison with medical therapy

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ABSTRACT

Background: To compare the outcomes in a group of patients with Budd-Chiari syndrome (BCS) managed by percutaneous recanalization with a group of patients who were managed by medical therapy alone.

Methods: We retrieved the hospital records of 37 patients with BCS admitted to our facility between 2004 to 2017 and identified 24 patients (male:female = 10:14; mean age, 32.7 ± 12.5 years) who underwent percutaneous recanalization. Remaining thirteen patients (male:female = 3:10; mean age, 36.77 ± 14.71 years), were managed by medical therapy. Technical and clinical results, complications, and primary patency of percutaneous recanalization were analyzed. Overall and symptom-free survival rates, the frequency of symptom recurrence, and the number of readmissions for recurrent symptoms were analyzed in both interventional treatment and medical therapy groups.

Results: Technical success for recanalization of hepatic vein/inferior vena cava by angioplasty $\pm$ stenting was achieved in 22 patients (22/24, 91.7%). Clinical success was achieved in 19 patients (19/24, 79.2%). Overall survival for patients who underwent percutaneous recanalization at 1 year and five years was 87.0% and 87.0% and for patients with medical therapy was 90.1% and 45.5%, respectively ($P = 0.710$). Symptom-free survival for patients who underwent percutaneous recanalization at 1 year and five year was 93.3% and 81.7% and for patients with medical therapy was 26.0% and 0%, respectively ($P < 0.001$). In the intervention group, 4 patients (4/24, 16.7%) were admitted for recurrent symptoms (median number of readmissions 1, range: 1–2) whereas in medically managed patients 9 patients (9/13, 69.2%) were readmitted (median number of readmissions, 2; range, 1–5) ($P = 0.003$).

Conclusion: There was no statistically significant difference in overall survival of patients managed with percutaneous recanalization and medical therapy. Percutaneous recanalization had definite benefit in terms of fewer recurrent symptoms and hospital admissions, hence should be performed whenever technically feasible.

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Keywords: Angioplasty; Ascites; Budd-Chiari syndrome; Myeloproliferative disorders; Thrombosis

Introduction

Budd-Chiari syndrome (BCS) results from hepatic venous outflow obstruction occurring anywhere from the level of the small hepatic veins (HVs) to the level of the termination of the inferior vena cava (IVC) into the right atrium in the absence of cardiac disease, pericardial disease, or sinusoidal obstruction syndrome.¹ In the European population, the underlying disorder leading to BCS is prothrombotic, whereas in the Asian population most patients have been reported to have a web/stenosis with no underlying

prothrombotic disorder.² No controlled trials have been performed because of the rarity of this disease. The current recommendations for the treatment of primary BCS originate from cohort studies and expert opinions. In Europe, a stepwise therapeutic strategy including anticoagulation, angioplasty/thrombolysis, transjugular intrahepatic portosystemic shunt (TIPS) and orthotopic liver transplantation is widely employed.³ However, in Asia, the majority of patients are treated directly by percutaneous recanalization of HVs or TIPS or with medical therapy.⁴ In this study, we evaluated patterns of obstruction, aetiological spectrum, technique and the

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outcome of percutaneous interventions, compared to outcome in a cohort of patients who were managed by medical therapy alone.

Methods

Institutional review board approval was taken for this retrospective analysis (Amrita Institute of Medical Sciences and Research Centre [AIMS], Kochi, India). We retrieved the hospital records of 37 patients with BCS admitted to our facility between 2004 to 2017 and identified 24 patients who underwent percutaneous recanalization. The remaining 13 patients, not feasible for recanalization were managed by medical therapy. The patients with malignancy other than myeloproliferative neoplasm were excluded. Clinical records and images were retrieved from electronic medical record and picture archiving and communication system.

Baseline characteristics of 37 patients with BCS are presented in Table 1, including clinical features, laboratory parameters, type of BCS, model for end-stage liver disease score (MELD), and BCS related prognostic index (Rotterdam score). The Rotterdam score was calculated using the following equation: $1.27 \times \text{encephalopathy} + 1.04 \times \text{ascites} + 0.72 \times \text{prothrombin time} + 0.004 \times \text{bilirubin}$. Ascites and hepatic encephalopathy were scored as present (1) or absent (0) and prothrombin time as higher (1) or lower (0) than 2.3 international normalized ratio (INR). Bilirubin was included as a continuous variable for which the risk was scored as 0.004 per $\mu\text{mol/L}$. The total score ranged from 0.02 to 4.03. Class 1 represented total score between 0 to 1.1, class 2 between 1.1 to 1.5 and class 3 score was 1.5 and higher.⁵

Statistical analysis

Statistical analysis was performed using IBM SPSS Statistics 20.0 software (IBM Co., Armonk, NY, USA). The data was analyzed using descriptive statistical methods. Continuous data between groups were compared using Mann-Whitney U test and categorical data was compared using Fisher's exact test. Statistical significance was established at a *P*-value of less than 0.05. Symptom-free and overall survival analysis was carried out using Kaplan-Meier curve and compared using log-rank test.

Definition and diagnosis

BCS was diagnosed using a combination of colour Doppler, computed tomography and magnetic resonance imaging, when there was obstruction of the inferior vena cava (IVC) with patent HV (IVC type), obstruction of any of the three main HVs (HV type) or obstruction of both IVC and any of three main HVs (combined type).^{1,6} The presence of ascites, pain abdomen, encephalopathy or jaundice was considered symptomatic BCS. All symptomatic patients were considered for percutaneous recanalization. Patients with technical contraindications to percutaneous recanalization, diffuse HV thrombosis and those not willing for interventions were managed with medical therapy alone.

Intervention technique

The treatment strategy was based on the level of obstruction.

For IVC-type BCS, initial IVC angiogram was performed from a transfemoral approach. A hydrophilic guide wire (Terumo, Tokyo, Japan) supported by a multipurpose catheter was used to cross the occluded segment of the IVC. In one patient where the lesion could not be crossed from a transfemoral approach, the

Table 1 Baseline Characteristics of Patients with Budd-Chiari Syndrome Undergoing Percutaneous Recanalization or Medical Therapy

Variable	Intervention (n = 24)	Medical therapy (n = 13)	<i>P</i> -value
Sex			0.301
Male:female	10:14	3:10	
Age (yr)	32.7 ± 12.5	36.77 ± 14.71	0.471
Symptom			0.071
Ascites	19	11	
Ascites + pain	4	0	
Malena	1	0	
Pain	0	2	
Etiology			0.030
Antiphospholipid antibody syndrome	5 (20.8)	3 (23.1)	
Myeloproliferative disorder	3 (12.5)	4 (30.8)	
No prothrombotic disease	16 (66.7)	3 (23.1)	
Homocystenuria	0	1 (7.7)	
Hypereosinophilic syndrome	0	1 (7.7)	
Hereditary thrombophilia	0	1 (7.7)	
Rotterdam class			0.401
Class 1 (0–1.1)	11 (45.8)	3 (23.1)	
Class 2 (1.1–1.5)	10 (41.7)	8 (61.5)	
Class 3 (1.5 and higher)	3 (12.5)	2 (15.4)	
Pattern of involvement			0.062
Hepatic venous	12 (50.0)	2 (15.4)	
Inferior vena cava	3 (12.5)	1 (7.7)	
Combined	9 (37.5)	10 (76.9)	
Laboratory parameters			
Total bilirubin ($\mu\text{mol/L}$)	41.4 ± 40.5	46.9 ± 25.9	0.151
Total bilirubin (mg/dL)	2.4 ± 2.3	2.7 ± 1.5	0.151
Creatinine ($\mu\text{mol/L}$)	88.1 ± 32.4	74.7 ± 9.3	0.205
Creatinine (mg/dL)	1.0 ± 0.4	0.8 ± 0.1	0.205
Prothrombin time (sec)	20.1 ± 5.4	21.3 ± 7.9	0.322
International normalized ratio	1.5 ± 0.4	1.8 ± 0.5	0.033
Albumin (mg/dL)	2.9 ± 0.8	3.4 ± 0.8	0.141
Aspartate transaminase (IU/L)	39.8 ± 21.1	92.5 ± 174.8	0.314
Alanine transaminase (IU/L)	30.8 ± 21.2	75.9 ± 16.7	0.539
Alkaline phosphatase (IU/L)	157.0 ± 135.3	170.7 ± 66.7	0.124
Platelet count ($\times 10^9/\text{L}$)	199.5 ± 133.5	317.4 ± 156.2	0.02
Sodium (mmol/L)	132.8 ± 6.3	134.6 ± 4.4	0.539
MELD score	13.9 ± 4.7	16.5 ± 3.6	0.064
Rotterdam score (0.02–4.03)	1.24 ± 0.43	1.17 ± 0.54	0.422

Values are presented as number only, mean ± standard deviation, or number (%).

MELD, model for end-stage liver disease.

wire was exchanged for a Rosch-Uchida transjugular liver access set (Cook Inc., Bloomington, IN, USA) and the inner trocar-catheter combination was used to traverse the occluded segment. The diameter of the balloons used for IVC dilation ranged from 8

Fig. 1. A 55-year-old female patient presenting with ascites. Model for end-stage liver disease (MELD) score of 16 and Rotterdam score of 1.16 (Class 2). Web-like occlusion in the suprahepatic inferior vena cava (IVC) (arrow; A). Right and middle hepatic veins were draining into the IVC distal to occlusion (not in image). Left hepatic vein was draining into IVC below occlusion (arrowhead; B). Through transfemoral approach the Rosch-Utschida set was positioned with the tip at the level of the web (arrow; C). The web was punctured by the inner trocar-catheter combination (arrow; D) and crossed (arrow; E). Note the contrast in the right atrium (arrow; E). Angioplasty performed using an 18mm balloon (arrow; F). Post-angioplasty angiogram shows the free flow of contrast across previous occluded segment (arrow; G) with no pressure gradient.

to 18 mm (Atlas; Bard Peripheral Vascular, Tempe, AZ, USA) (Fig. 1). The IVC stent used was 14 mm in diameter (E-Luminex; Bard Peripheral Vascular).

For HV-type BCS, the HV recanalization was performed via transjugular or transhepatic route. Involvement of the ostium of the vein makes access from the transjugular route difficult. If the HV proximal to occlusion was patent and of adequate size, it was targeted for percutaneous recanalization. The target HV was accessed under ultrasound guidance using a Neff set (Cook Inc.) and a 6 Fr sheath was placed in the vein. The occlusion or stenosis was then crossed with a hydrophilic wire (Terumo) supported by a Kumpe catheter (Cook Inc.). In one patient the wire was snared from a right internal jugular vein access and recanalization was performed from above. In 23 patients, the procedure was performed through a transhepatic percutaneously placed 6 Fr sheath. Balloon expandable stents of 8 to 10 mm diameter and 37 to 57 mm length (Bard Peripheral Vascular or Express Biliary stent [Boston Scientific, Marlborough, MA, USA] or Scuba [Medtronic, Dublin, Ireland] or Omnilink Elite [Abbott Vascular, Santa Clara, CA, USA]) were placed in the patients with focal stenosis or short segment occlusion (< 3 cm). In patients with > 3 cm long occlusion or stenosis, a self-expandable metal stent was preferred. In one patient with left HV (LHV) stenosis angioplasty alone was performed for financial reasons (Fig. 2).

In the combined type, initially the IVC stenosis was assessed for pressure gradient via transfemoral/transjugular approach, if significant (> 10 mmHg), angioplasty was performed and then the accessible target HV was recanalized by angioplasty and stenting via transjugular/transhepatic approach.

Following radiological intervention all these patients were put on a strict anticoagulation regime with the aim of keeping INR in

the range of 2 to 3. The patients who did not undergo radiological intervention were also anticoagulated to keep INR in the range of 2 to 3, in addition to symptomatic therapy (low-salt diet, diuretic therapy and/or beta-blockers as needed) and chemotherapy wherever appropriate. The INR range of 2 to 3 was decided according to European Association for the Study of the Liver guidelines.⁷

Study endpoints

Technical success, clinical success, primary patency, and complications of percutaneous recanalization were analyzed. Overall and symptom-free survival rates, the frequency of symptom recurrence, and the number of readmissions for recurrent symptoms were analyzed in both interventional treatment and medical therapy groups.

Technical success was defined as restoration of flow across the occluded segment in the immediate post-procedure angiography. Inability to restore the flow across the occluded segment was defined as technical failure. Clinical success was defined as improvement or stabilization of patient's symptoms or liver function after technically successful recanalization of hepatic venous outflow.⁸ Clinical failure was considered when there was no improvement in patient's symptoms, the onset of new symptoms or deterioration of liver function. Primary patency was defined as the time interval from technically successful procedure to the detection of recurrent symptoms, reocclusion on imaging or the last follow-up. A recurrence was considered to have occurred if more than 50% narrowing of the treated segment was seen at imaging done after completion of treatment. Symptom-free survival was defined as the time interval from technically successful procedure to the detection of recurrent symptoms or death. Overall survival was

Fig. 2. A 49-year-old male patient with polycythemia vera presenting with ascites. Model for end-stage liver disease (MELD) score of 13 and Rotterdam score of 1.14 (Class 2). Short segment tight stenosis involving left hepatic vein ostium (arrow; A). Right hepatic vein and middle hepatic vein were occluded. Inferior vena cava was patent. Through transhepatic approach stenosis crossed (arrow; B) and 8 mm × 37 mm balloon expandable stent deployed across the stenotic segment (arrow; C). Post-stenting angiogram shows the free flow of contrast across the stent (arrow; D).

defined as the time from start of treatment to time until death, the end of the study period or the date of the last visit. Follow-up was done by imaging (ultrasound, computed tomography, magnetic resonance imaging) and clinical assessment.

Complications were classified as major and minor according to the guidelines of the Society of Interventional Radiology Standards of Practice Committee.⁹

Results

Most baseline patient characteristics, except etiology, platelet count, and INR, were similar between intervention and medical therapy groups (Table 1). Ascites was the most common symptom in both groups (79.2% in intervention group and 84.6% in medical therapy group). As for etiology, 8 of 24 patients (33.3%) in intervention group and 10 of 13 patients (76.9%) in medical therapy group had the underlying systemic prothrombotic disorder ($P = 0.030$). The platelet count and INR in medical therapy group were significantly higher than the intervention group ($P = 0.020$ for platelet count and $P = 0.033$ for INR). The mean Rotterdam score in the intervention group (1.24 ± 0.43) was higher than in medical therapy group (1.17 ± 0.54); however, this was not statistically significant ($P = 0.422$).

Intervention

In patients with HV or HV + IVC pattern of BCS, all three HVs were partially or completely occluded and the vein with the shorter length of occlusion or stenosis was chosen as the target for recanalization. Out of 12 patients (50.0%) with HV pattern of involvement, technical success was achieved in 11 patients (91.7%). Ten target HVs (LHV-4, middle HV [MHV]-2, right HV [RHV]-4) were recanalized with stenting and one LHV occlusion was recanalized by angioplasty alone. Technical success was achieved in 8 out of 9 patients (88.8%) with combined HV + IVC pattern (Fig. 3). In three of these patients, IVC angioplasty was performed initially followed by recanalization of HVs (MHV-2, LHV-1) by stenting. One IVC could not be recanalized and only the LHV was stented. In two patients, the IVC stenosis was not hemodynamically significant and hence only the LHV and MHV respectively were treated with stents. In two patients, only IVC angioplasty was performed as the HVs were not amenable to the treatment. In one patient, transhepatic approach to recanalize the RHV failed and subsequently patient underwent portocaval shunt. In the three patients with pure IVC pattern of involvement, one was treated successfully by angioplasty alone and the other two by stenting. Overall technical success was achieved in 22 patients (22/24, 91.7%). The primary patency at 1, 3, and 5 year was 93.3%, 81.7%, and 81.7%, respectively (Fig. 4). In a patient during an attempt to recanalize the MHV through transhepatic approach,

Fig. 3. A 30-year-old male patient presenting with ascites. Model for end-stage liver disease (MELD) score of 16 and Rotterdam score of 1.4 (Class 2). Short segment occlusion involving right hepatic vein (RHV) ostium (arrow; A). Middle hepatic vein and left hepatic vein were occluded. Retrohepatic inferior vena cava (IVC) stenosed for short segment (arrow; E). Through transhepatic approach, the RHV occlusion crossed (arrow; B) and 8 mm × 37 mm balloon expandable stent deployed across the occluded segment (arrow; C). Post-stenting angiogram shows the free flow of contrast across the stent (arrow; D). The IVC stenosis was crossed through the transfemoral approach and angioplasty performed using 18 mm balloon (arrow; F). Post-plasty angiogram shows the free flow of contrast across the previous stenotic segment (arrow; G).

the guidewire punctured the HV and entered the right atrium directly without entering the IVC. Angioplasty resulted in massive pericardial tamponade. Despite performing emergency thoracotomy and repair of the tear in the myocardium the patient could not be saved (Fig. 5). In another patient who underwent LHV stenting, the stent thrombosed two days after the procedure and patient died within a week. In a post liver transplant patient, thrombosis occurred immediately after procedure following which, thrombolysis was attempted. However, patient died due to graft failure. Intention to treat analysis showed the clinical success in 19 patients (19/24, 79.2%). Delayed stent thrombosis was seen in 2 patients

(2/22, 9.1%). In one patient, thrombosis occurred at 63 months. A patient who underwent RHV stenting, developed thrombosis of the stent at 26 months because anticoagulation was stopped during pregnancy. This patient was successfully managed after child birth by thrombolysis followed by angioplasty and stenting and the follow-up Doppler at 8 months showed patent stent (Fig. 6).

In patients who underwent percutaneous recanalization, post-treatment follow-up period was 37.5 ± 33.2 months (range, 3–108 months) and was available in all patients who successfully underwent recanalization. Applying Kaplan-Meier curves, the mean survival time was 93.9 months (95% confidence interval [CI], 79.1–

108.8 months). In patients with medical therapy, post-treatment follow-up period was 27.5 ± 30.9 months (range, 1–96 months; median, 9 months) and was available in all patients. The mean

Fig. 4. Kaplan-Meier curve for primary patency in patients with Budd-Chiari syndrome managed by percutaneous recanalization.

survival time was 77.25 months (95% CI, 57.42–97.07 months). For patients who underwent percutaneous recanalization, overall survival at 1 year was 87.0% and at five years was 87.0% and for patients managed by medical therapy, overall survival at 1 year was 90.1% and at five years was 45.5% ($P = 0.710$, log-rank test; Fig. 7). The symptom-free survival at 1 year and 5 years for the patients who underwent percutaneous recanalization was 93.3% and 81.7%, respectively and for patients managed by medical therapy, it was 26.0% and 0%, respectively ($P < 0.001$, log-rank test; Fig. 8). We analyzed the number of readmissions for recurrent ascites, pain or gastrointestinal bleed in both the groups. In the intervention group, 4 patients (4/24, 16.7%) were readmitted whereas in medically managed patients 9 patients (9/13, 69.2%) were readmitted ($P < 0.001$). The median number of readmissions of patients managed by percutaneous recanalization was 1 (range, 1–2), whereas in medically managed patients median number of readmissions was 2 (range, 1–5) ($P = 0.003$). The patients who underwent percutaneous recanalizations had a fewer recurrence of symptoms and readmissions in the follow-up period (Table 2).

Discussion

The pattern of hepatic venous obstruction varies in different parts of the world. The literature suggests that IVC obstruction

Fig. 5. A 15-year-old female patient presenting with abdominal pain and ascites. Model for end-stage liver disease (MELD) score of 10 and Rotterdam score of 1.1 (Class 1). The hepatic venogram showed stenosis involving distal middle hepatic vein (arrows; A, B). Right hepatic vein and left hepatic vein were occluded. Inferior vena cava (IVC) was patent. Through transhepatic approach, multiple attempts were made to cross the occlusion. The guide wire punctured the hepatic vein and entered the right atrium directly (arrow; C) without entering the IVC, which was missed initially. Angioplasty (arrow; D) resulted in massive pericardial tamponade (arrow; E). Despite performing emergency thoracotomy and repair of the tear in the myocardium the patient could not be saved.

Fig. 6. A 25-year-old female patient with polycythemia vera (JAK 2 mutation) presenting with ascites. Model for end-stage liver disease (MELD) score of 16 and Rotterdam score of 1.23 (Class 2). Short segment occlusion involving right hepatic vein ostium (arrow; A). Middle hepatic vein and left hepatic vein were occluded. Inferior vena cava was patent. Through transhepatic approach occlusion crossed (arrow; B) and 8 mm × 37 mm balloon expandable stent deployed across the occluded segment (arrow; C). Post-stenting angiogram shows the free flow of contrast across the stent (arrow; C). Stent thrombosis occurred at 26 months, which was successfully treated by thrombolysis, angioplasty, and stenting. Follow-up Doppler at 8 months showed patent stent (not shown in image).

Fig. 7. Kaplan-Meier curves for overall survival in patients with Budd-Chiari syndrome managed by percutaneous recanalization and medical therapy. Overall survival: $P = 0.710$, log-rank test (percutaneous recanalization: 1 yr, 87.0%; 5 yr, 87.0%; medical therapy: 1 yr, 90.1%; 5 yr, 45.5%).

is commoner in the East while HV thrombosis is commoner in the West.² However recent studies from India suggests that combined involvement of HV and IVC is commoner than previously observed.^{10–13} The decrease in the proportion of pure IVC involvement can very well be due to the availability of better non-invasive imaging, allowing recognition of the associated HV obstruction and differentiation of collaterals from HV. In our study population, we found that combined HV and IVC obstruction as the commonest pattern (51.4%) (Table 1). The previous studies have suggested that in Asia, BCS results from web/stenotic lesions in the IVC/HV whereas in the West an underlying prothrombotic disorder causes HV thrombosis.² In our study, 18 patients (18/37, 48.6%) had underlying prothrombotic conditions (Table 1). The high percentage of underlying prothrombotic disorders may be a direct result of the easy availability of molecular tests to detect these abnormalities. Studies have shown that long-standing thrombosis can lead to the formation of web/membrane like lesions.¹⁴ In the patients in whom we performed an intervention, we were able to identify an underlying prothrombotic condition in only 33.3%, whereas in patients managed by medical therapy alone, 76.9% of patients had an underlying prothrombotic condition ($P = 0.030$; Table 1). The patients in medical therapy group had significantly higher platelet count and INR, compared to the intervention group. The Rotterdam score showed no statistically

Fig. 8. Kaplan-Meier curve for symptom-free survival in patients with Budd-Chiari syndrome managed by percutaneous recanalization and medical therapy. Symptom-free survival: $P < 0.0001$, log-rank test (percutaneous recanalization: 1 yr, 93.3%; 5 yr, 81.7%; medical therapy: 1 yr, 26.0%; 5 yr, 0%).

significant difference between two groups. However, the scoring systems do not take into consideration the pattern and extent of the vascular involvement. These factors could have had a bearing on the outcome of the study. Recent studies from Asia have shown the effectiveness of percutaneous recanalization in various BCS patients and the survival rates in these patients were comparable to that of a European population undergoing TIPS and orthotopic liver transplantation.^{4,15–21} In our study we achieved technical success of 91.7% for HV pattern, 100% for IVC pattern and 88.8% for combined HV and IVC pattern of BCS. Clinical success was achieved in 79.2% of patients.

The subset of patients in whom HV/IVC recanalization was not performed, were managed by only medical therapy to relieve symptoms and treatment of the underlying prothrombotic disorder if detected. We did not attempt TIPS in any of these patients due to both technical and financial reasons. Performing TIPS in these patients is challenging as there is no patent HV available for the creation of the shunt.

Medical therapy consists of anticoagulation, sodium restriction, diuretic therapy and drugs for treating the underlying cause. Zeitoun et al²² compared the survival of patients managed by

Table 2 Outcome Data of Patients with Budd-Chiari Syndrome Managed by Percutaneous Recanalization or Medical Therapy

Outcome	Percutaneous recanalization	Medical therapy	P-value
Overall survival (%)			0.710
1 yr	87.0	90.1	
5 yr	87.0	45.5	
Symptom-free survival (%)			<0.001
1 yr	93.3	26.0	
5 yr	81.7	0	
Recurrence	4/24 (16.7)	9/13 (69.2)	<0.001
No. of readmissions	1 (1–2)	2 (1–5)	0.003

Values are presented as %, number (%), or median (range).

surgical shunts, with patients managed only by medical therapy. They observed no statistically significant and independent influence of surgical portosystemic shunts on survival. They concluded that overall improved survival was due to the improved management of hypercoagulable states as well as treatment of the underlying cause. In a multi-center prospective study by Darwish Murad et al,²³ 80 patients (80/163, 49.1%) were managed without any invasive intervention and 13 patients (13/163, 8.0%) received no treatment. They observed excellent short-term survival rates, sometimes better than those observed in earlier retrospective series, in which more interventions (such as surgical shunting) were used.^{3,21} In our study, 13 patients (13/37, 35.1%) were managed by medical therapy alone. These patients were in an advanced stage of the disease (average Rotterdam score 1.17 ± 0.54 ; 61.5%, class 2; 23.1%, class 1; 15.4%, class 3). An underlying prothrombotic disorder was detected in 76.9% of the patients. No significant difference was seen in overall survival between patients managed by intervention and by medical therapy. The fact that in 76.9% of the patients, the underlying cause was detected and treated might have influenced the overall outcome. Overall five-year survival was less (45.5%), though not statistically significant when compared to the intervention group (87.0%). We also found that for patients who were managed by medical therapy alone, the symptom-free survival at 1 year and 5 years was 26.0% and 0%, respectively which was significantly less compared to patients who underwent percutaneous recanalization, for whom it was 93.3% and 81.7%, respectively. Moreover, these patients required more frequent hospital admissions to manage the recurrent symptoms (ascites, gastrointestinal bleed) compared to the intervention group.

Our series, like many studies, is small. TIPS, HV/IVC recanalization and medical therapy alone compared to each other in a larger cohort of patients could provide useful information for the decision making in this challenging disease.

The key observation from our study is that there is no statistically significant difference in overall survival of patients managed with percutaneous recanalization and medical therapy. Percutaneous recanalization had definite benefit in terms of fewer recurrent symptoms and hospital admissions, hence should be performed whenever technically feasible.

Conflicts of Interest

No potential conflict of interest relevant to this article was re-

ported.

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