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Percutaneous transhepatic venoplasty: an alternative treatment for Budd-Chiari syndrome

Perkütan transhepatik venoplasti: Budd-Chiari sendromu için alternatif bir tedavi

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Background/aims: *In patients with Budd-Chiari sydrome due to short segment hepatic vein stenosis where percutaenous transluminal venoplasty is not successful, percutaneous transhepatic balloon venoplasty may be a valid treatment option. The aim of this prospective study was to evaluate the effects of this procedure for the treatment of patients with Budd-Chiari sydrome, in whom transluminal cannulation was unsuccessful.* ***Methods:*** *Ten patients with short segment occlusion of the hepatic veins were treated by percutaneous transhepatic balloon venoplasty between January 1997 and January 2000. The median follow-up period of these patients was 20 months (2-33 months).* ***Results:*** *The procedure was unsuccessful in two patients. Eight patients (five men, three women) with a median age of 28 (range, 15-61) years were treated by percutaneous transhepatic balloon venoplasty and in seven of them, clinical symptoms including abdominal distension and ascites, resolved completely. Long term anticoagulation therapy was not given to the patients. One patient with advanced stage liver disease died of variceal bleeding two months after the procedure. During follow-up, symptomatic reocclusion requiring dilatation occurred in three patients.* ***Conclusions:*** *Percutaneous transhepatic balloon venoplasty is an alternative treatment option for selected patients with Budd-Chiari sydrome when transluminal cannulation of the hepatic veins is not possible. Long term anticoagulation therapy seems to be necessary in these patients.*

Key words: Budd-Chiari syndrome, short segment hepatic venous stenosis, percutaneous transhepatic balloon venoplasty.

Amaç: *Kısa segment hepatik ven tıkanmasına bağlı olarak gelişen ve perkutan transfemoral venoplatinin başarısız olduğu Budd-Chiari sendromlu vakalarda perkutanöz transhepatik balon venoplasti geçerli bir tedavi seçeneği olabilir. Bu çalışmanın amacı transluminal kanulasyonun başarısız olduğu Budd-Chiari sendromlu'lu hastaların tedavisinde perkutanöz transhepatik balon venoplasti'nin etkinliğini araştırmaktı.* ***Yöntem:*** *Ocak 1997 ve Ocak 2000 yılları arasında kısa segment hepatik ven tıkanması olan 10 hasta PTBV ile tedavi edildi. Hastaların ortalama takip süresi 20 (2-33 ay arası) aydı.* ***Bulgular:*** *İki hastada işlem başarısızdı. Ortanca yaşları 28 olan (15- 65 yaş arası) 8 hasta (5 erkek, 3 kadın) PTBV ile tedavi edildi. Yedi hastada karında gerginlik ve asit tamamen kayboldu. Uzun süreli antikoagulasyon hastalara uygulanmadı. Son dönem karaciğer hastalığı olan bir hasta işlemden iki ay varis kanaması nedeniyle vefat etti. Takip süresince tekrar dilatasyonu gerektiren semptomatik damar tıkanması tekrarı 3 hastada görüldü.* ***Sonuç:*** *Perkutanöz transhepatik balon venoplasti seçilen bazı Budd-Chiari sendromlu'lu hastalarda transluminal yolla kanulasyonun mümkün olmadığı durumlarda alternatif bir tedavi yöntemi olabilir. Uzun süreli antikoagulasyon gerekli gibi görünüyor.*

Anahtar kelimeler: Budd-Chiari sendromu, kısa segment hepatik ven tıkanması, perkutanöz transhepatik balon venoplasti.

INTRODUCTION

Budd-Chiari syndrome (BCS) is an uncommon, often fatal disorder caused by the occlusion of major hepatic veins, the suprahepatic inferior vena cava (IVC), or both (1,2). Since the site, extent and rapidity of hepatic vein occlusion is highly variable, there is a range of clinical presentations necessitating an individualized therapeutic strategy in each patient (3,4). Medical management of BCS has been generally unsatisfactory (5). Several published series have advocated surgical portosysthemic shunting as an appropriate option for patients with stable liver function (1,3,4), with liver transplantation reserved for those presenting with fulminant liver failure or end stage liver disease (1,6). Percutaneous transluminal balloon venoplasty (PTBV) has been used

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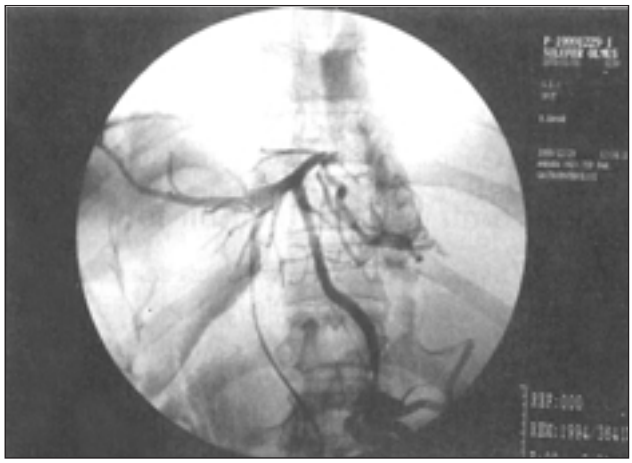

Figure 1. Percutenous hepatic angiography via the xyphoid approach demonstrating occlusion at the confluence of the left hepatic vein and the inferior vena cava

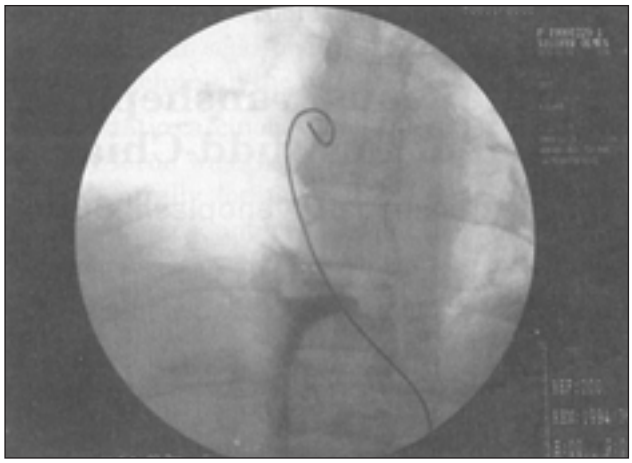

Figure 2. A guide wire is passed from the left hepatic vein into the vena cava inferior.

for treatment of BCS due to webs of the suprahepatic portion of the IVC and short segment hepatic vein occlusion (7-9). Combined transhepatic and transjugular hepatic balloon venoplasty may also be considered for the treatment of patients with BCS caused by short length hepatic venous occlusion (3,10). In patients in whom transluminal venoplasty can not be achieved, successful hepatic balloon venoplasty restores physiologic hepatic venous drainage and may thus be pathophysiologically preferred to surgical shunting.

In this study we report our experience with PTBV in a prospectively assessed patient population.

MATERIALS AND METHODS

Ten patients (six men, four women), who were admitted to our department with the diagnosis of BCS between January 1997 and January 2000, were prospectively studied. The diagnosis of BCS was confirmed by imaging techniques including Doppler ultrasound examination and hepatic venography and cavography by the transfemoral route as well as liver biopsy when required. In every case, all known possible etiologic causes of BCS were investigated before therapeutic intervention. Patients were included in the study according to the following criteria: 1. Short segment (less than 2 cm in diameter) hepatic venous occlusion with or without suprahepatic IVC occlusion, 2. No previous surgical shunting, 3. Patent segment above occluded segment of at least one hepatic vein, 4. Unsuccessful hepatic vein cannulation by transluminal route. 5. No presence of coagulopathy impending transhepatic intervention, Patients with BCS caused by primary or metastatic malignancy were excluded from the study.

The study was approved by the University of Ankara Medical School Ethics Committee and all patients gave informed consent to participation.

Clinical follow-up and response to therapy

Comprehensive clinical history, complete physical examination, serum biochemical values, complete blood count and the findings of abdominal ultrasound were recorded for all patients before therapeutic intervention and after six months or when symptoms recurred. Esophagogastroduodenoscopy was performed when required.

Therapeutic intervention

After sedation, local and general analgesia, transhepatic venography was performed with real-time sonographic guidance and flouroscopic control according to a previously described method (11). During hepatic venography the occluded vascular segment was identified and dilatation of this occluded segment was performed using guidewire, a 6-F sheath, and a polyethylene balloon varying from 8-12 mm in diameter and from 5-9 cm in length (Boston Smast balloon, Ultra-thin balloon, Bluemak balloon, Tanintraduter sheat, Boston sheat) according to the previously described method (11). At initial inflation, the occluded short segment was easily recognizable by a localized narrowing of the hepatic vein or IVC at

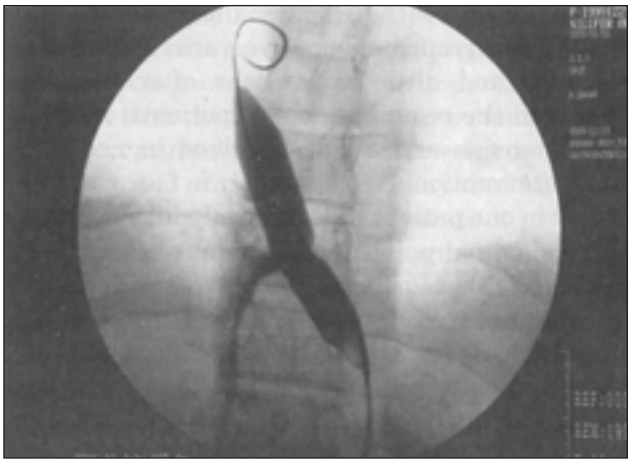

Figure 3. A baloon is passed over the guide wire. The inflated baloon clearly displays the narrowed segment at the left hepatic vein- inferior vena confluence.

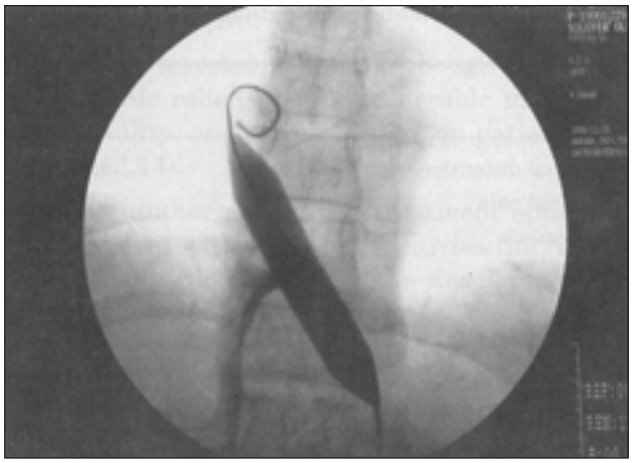

Figure 4. With the baloon catheter fully inflated normal venous caliber is established.

angiography. Inflation pressure was continued until this narrowing disappeared and good antegrade flow was achieved (Figure 1-4). The sheath and guide-wire was then withdrawn and percutaneous compression was applied to the insertion point for 30 minutes to achieve haemostasis. Patients were monitored for 24-48 hours in the intensive care unit for possible procedure related complications. Anticoagulation therapy with low-molecular weight heparin (fraxiparin, 0.3 ml, subcutaneously b.i.d), was prescribed to every patient for a period of one month. Oral anticoagulants were not prescribed at any time after the intervention.

RESULTS

The procedure was successful in eight patients. In two patients, the occluded segment of the hepatic veins was too long and it was not possible to traverse this segment with a guide-wire. Eight patients (five men, three women) with a median age of 28 (range, 17-61) years were treated with PTBV. The median duration of disease prior to therapeutic intervention was 17 (range, 5-60) months. In two cases, BCS could have been related to polycytemia rubra vera while in the other six cases, the cause could not be identified.

Angiography findings

Table 1 summarizes the angiography findings of patients in whom successful dilatation was achieved by PTBV. One patient had only membranous occlusion at the confluens of the hepatic veins and the IVC. In the remaining seven patients, short segment thrombotic occlusion of one or two hepatic veins was observed. One of these seven patients also had a membranous IVC obstruction.

Table 1. The findings of cavography and transhepatic hepatic venography of patients with successful dilatation.

No	*RHV*	*MHV*	*LHV*	*IVC*	*Cause*	*Restenosis*
1	Dilated	Occluded	Dilated	Occluded	Membrane	24 months
2	Occluded	Dilated	Occluded	Patent	Thrombus	18 months
3	Dilated	Occluded	Occluded	Patent	Thrombus	No
4	Occluded	Occluded	Dilated	Patent	Thrombus	No
5	Occluded	Dilated	Occluded	Thrombosis	Thrombus	No
6	Dilated	Occluded	Occluded	Patent	Thrombus	No
7	Occluded	Dilated	Occluded	Patent	Thrombus	8 Months
8*	Dilated	Dilated	Dilated	Patent	Membrane	No

RHV: right hepatic vein, MHV: medial hepatic vein, LHV: left hepatic vein,
IVC: inferior vena cava, *: Membrane between confluence of hepatic veins and IVC.

Table 2. Clinical features of patients before and after PTBV.

Parameter	*Before PTBV N (%)*	*After PTVB N(%)**
Abdominal distension	8(100)	1(12.5)
Abdominal pain	2 (25)	1 (12.5)
Hematemesis	1 (12.5)	1 (12.5)
Hepatomegaly	8 (100)	4 (50)
Ascites	8 (100)	1 (12.5)
Splenomegaly	4 (50)	3 (37.5)
Varices	3 (37.5)	2 (25)
Edema	2 (25)	1 (12.5)

PTBV: Percutaneous transhepatic balloon venoplasty
*: until the second PTBV or last clinical visit.

Dilatation was performed through the left hepatic lobe in five patients and through the right hepatic lobe in three patients. A balloon catheter of 8 mm diameter was used in seven patients, and in one patient, dilatation of the narrowed segment was achieved with a 12 mm balloon. In one patient with complete membranous occlusion in the IVC, transluminal dilatation was performed as described previously (12) prior to PTBV. After this intervention, edema in the lower extremeties disappeared. However, no effect on ascites and hepatomegaly was seen, and seven months after transluminal cavoplastiy PTBV was therefore performed.

Clinical features and follow-up

Pre- and postintervention clinical features of patients are summarized in Table 2. The most common symptom was abdominal distension and the most common pathologic finding on physical examination was hepatomegaly and ascites. Edema in lower extremeties was observed only in the two patients who had concomittant IVC occlusion. Esophagogastroduodenoscopy showed varices and portal gastropathy in three patients, of whom two had grade II varices and one had grade IV varices. One patient had a right sided pleural effusion causing dyspnea and necessitating thoracentesis.

The median follow-up after intervention was 20 (range, 2-33) months. In one patient, who had liver cirrhosis and portal hypertension, restenosis developed after 15 days and PTBV had to be repeated. A self expandable wall-stent was placed between the hepatic vein and IVC. Three days after the procedure, the patency of the wall-stent was verified with Doppler-ultrasound. This patient had recurrent massive variceal bleeding, however, and died two months after the first PTBV. In the remaining seven patients, abdominal distension and ascites resolved immediately after intervention. Hepatomegaly in four patients, edema in one patient, splenomegaly in one patient and abdominal pain in one patient also resolved. Apart from these patients, recurrence of occlusion occurred in three other patients during follow-up. The first had idiopathic BCS with a short segment thrombosis of the hepatic veins. The second had membranous occlusion in the suprahepatic IVC and at the level between the hepatic veins and IVC and the third patient had BCS caused by polycytemia rubra vera. These three patients had became asymptomatic after the first PTBV but their initial presenting symptoms recurred 24, 18 and 8 months after therapeutic intervention respectively. Repeated hepatic venography showed occlusion at the location of the primary occlusion site. PTBV was performed again with concomitant improvement of the patients' symptoms and physical findings.

Complications

All patients tolerated this procedure very well except for one patient who had slight temporary hypotension during procedure. There was no bleeding complication at the procedure site in any patient.

DISCUSSION

Our results confirm that PTBV is an alternative treatment option for selected patients with BCS in whom transluminal cannulation of the hepatic vein is not possible. The management of symptomatic patients with this disorder has traditionally consisted of conservative medical treatment, surgical shunting, radiologic intervention and orthotopic liver transplantation (1,6). Medical management alone is generally ineffective (2,5). Surgical porthosystemic shunting has been advocated as the most appropriate option for patients with stable liver function, with liver transplantation being reserved for those presenting with fulminant liver failure or end stage liver disease (13,14). The known radiologic interventions for the treatment of BCS include transluminal balloon dilatation of the IVC (5,7,9,14) and the hepatic veins (5,7), with and without stent placement (8) and transjugular intrahepatic portosysthemic stent-shunt (TIPS) (15,16). Hepatic vein

dilatation by the transhepatic route is another option which was preferred by the present authors although it is generally less often undertaken (11). We think that the transhepatic route enables easy and direct cannulation of hepatic veins by ultrasound –guided puncture. PTBV may thus be superior to transluminal angioplasty since it provides direct access to the occluded segment enabling easier manipulation of the guide-wire through the occlusion. The lack of success with the transluminal route in our cases supports this view. The only drawback may be that one is limited with respect to the size of the balloon catheter during the transhepatic procedure. We used a 8 mm balloon catheter in seven patients and a 12 mm balloon catheter in one patient.

PTBV can be performed safely when appropriate precautions are taken, which include the following: identifying a dilated and patent segment of hepatic vein, making a limited number of punctures, using the smallest catheter possible and using postpuncture compression to produce adequate haemostasis. In our patients, we were able to cannulate a hepatic vein using ultrasound guidance in all eight cases. In these eight cases with short segment BCS, dilatation was performed safely and restoration of hepatic blood flow was obtained during the procedure. Among these eight cases there was only one case without symptomatic improvement and who died of variceal bleeding two months after the procedure. Although the stent placed into the narrowed segment was patent by ultrasound three days after the procedure, the development of fatal variceal bleeding suggests restenosis, although this could not be documented. The length of the occluded segment seems to be critical for the success of PTBV. The lack of success in our two cases with long segment BCS support this view. Embolisation of the cannulation tract before catheter removal from the hepatic vein has been advocated to reduce the risk of bleeding from the intervention site (11). We did not embolize the cannulation tract but haemostasis was nevertheless achieved by carrying out compression alone at the cannulation site.

Surgical portosystemic shunts, though providing symptomatic relief, carry considerable morbidity and mortality, and have no effect on patient survival (4,13,14).

TIPS is an other alternative treatment option in selected cases with BCS, but it carries the risk of some complications such as the development of hepatic encephalopathy, decrease of blood supply to the liver parenchyma, mortality related to the intervention and a high rate of stent occlusion (17,18). Both surgical portosystemic shunt and TIPS can cause deterioration in liver parenchymal function by decreasing hepatopedal flow in small branches of the portal vein. In contrast, hepatic vein dilation, irrespective of the primary puncture site, improves physiologic hepatic blood flow and can be repeated easily when restenosis occurs. Hence, hepatic vein dilation should be preferred to shunt procedures in cases with short segment BCS.

We did not use routine oral anticoagulation therapy in line with the practice of some centers (5,9), but re-stenosis developed in half of our eight patients, which supports the view that short-length BCS is the sequela of localized thrombosis (19) and that full dose anticoagulation therapy may be necessary in these cases. However, in three of the four patients with re-stenosis, occlusion developed as late as 12, 14 and 18 months after PTBV. The relatively long term interval between the PTBV procedure and occlusion development may suggest the contribution of other factors as well.

In summary, PTBV is an alternative treatment option for BCS caused by short length segment occlusion of the hepatic vein, at least in cases where transluminal cannulation is not possible. At our institution, we have adopted this approach as the first line treatment in such cases. It should be performed before development of end stage liver disease. Larger series and longer follow-up periods are needed to determine the long term effects of PTBV on the course of BCS.

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