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Treatment of Budd-Chiari Syndrome by Metallic Stent as a Bridge to Liver Transplantation

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Abstract

A 49-year-old male with Budd-Chiari syndrome complicated by liver cirrhosis and intractable ascites is reported. The left hepatic vein was stenosed by a short subocclusive ostial web; the right and medial hepatic veins were thrombosed. A spontaneous intrahepatic portosystemic shunt had developed between the left portal and left hepatic veins. After ineffective balloon angioplasty, the left hepatic venous outflow was restored by placement of a 10-mm-diameter Wallstent across the web via a femoral approach. The hepatic venous pressure dropped from 29 to 12 mmHg. Rapid clinical improvement followed. The patient underwent liver transplantation 3 months later in stable condition.

Key words: Hepatic veins, thrombosis—Transluminal angioplasty—Stents—Budd-Chiari syndrome

Budd-Chiari syndrome is caused by thrombosis, long segmental stenosis, or membranous webs of the hepatic veins and/or the distal inferior vena cava [1]. In non-Oriental patients, extensive thrombosis of the venous hepatic outflow is common, whereas most cases of membranous webs are reported in patients from the Far East. Percutaneous transluminal angioplasty (PTA) has been described as an alternative to surgery, but may fail in 50% of nonselected patients after a 6-month follow-up [2]. The limited number of published cases treated with metal expandable stents prompted us to report this case and review the literature.

Case Report

A 49-year-old male was admitted with the diagnosis of liver cirrhosis complicated by ascites. Alcohol consumption was denied. Upper gastrointestinal endoscopy showed grade II gastric and esophageal varices. Beta blockers and diuretics improved the condition of the patient, who was readmitted 3 months later for recurrent ascites and edema of the lower limbs. Hepatic biopsy showed active micronodular cirrhosis. Diuretics were continued at a decreasing dose. Three months later the patient was operated on for a strangulated umbilical hernia. Ascites became intractable, respiratory function was impaired, and hepatic encephalopathy was noted. Gastroesophageal varices had enlarged and liver function had deteriorated with progressive cholestasis and hepatorenal syndrome.

Color-coded duplex (CCD) ultrasound demonstrated thrombosis of the medial and right hepatic veins and a tight ostial stenosis of the left hepatic vein. A long, tortuous, spontaneous intrahepatic portosystemic shunt, 1 cm in diameter, connected the left portal vein to the left hepatic vein. Flow was reversed in the right portal vein and arterial flow was increased. The inferior vena cava (IVC) was patent. Computed tomography (CT) confirmed CCD findings and major intrahepatic perfusion disturbances, mild splenomegaly, and perisplenic and left gastric varices. Celiac and superior mesenteric arteriography confirmed the intrahepatic portosystemic shunt and hepatofugal portal flow (Fig. 1). Phlebography by a right jugular approach confirmed thrombosis of the right and medial hepatic veins (Fig. 2). Opacification of the left hepatic vein failed due to the tight stenosis and the angle between hepatic vein and IVC. The concentric subocclusive ostial web was successfully catheterized by a right femoral approach (Fig. 3A). Pressure in the left hepatic vein measured 29 mmHg over 14 mmHg in the IVC. PTA of the web was performed with a 10-mm-diameter balloon, 4 cm in length (Meditech, Boston Scientific, Watertown, MA, USA), but the stenosis did not respond after balloon inflation to 17 atm for 2 min (Fig. 3B). A Wallstent (Schneider, Buelach, Switzerland), 10 mm in diameter and 4 cm in length, was placed at the level of the stenosis with a slight stent protrusion in the caval lumen. Normal venous drainage was restored (Fig. 3C). Hepatic venous pressure dropped to 12 mmHg. The patient was fully anticoagulated for 2 days. Diuretics were maintained for 2 weeks until liver tests normalized, except gamma-glutamyl transferase and alkaline phosphatase, which remained 2.5 times normal values. The overall clinical status improved rapidly. Three months later, liver transplantation was performed under stable conditions. The stent was removed without difficulty. The patient had experienced one minor episode of esophageal variceal bleeding, despite the spontaneous intrahepatic portosystemic shunt and a low portal pressure.

Discussion

Common causes of primary Budd-Chiari syndrome include hematological disorders, pregnancy, postpartum

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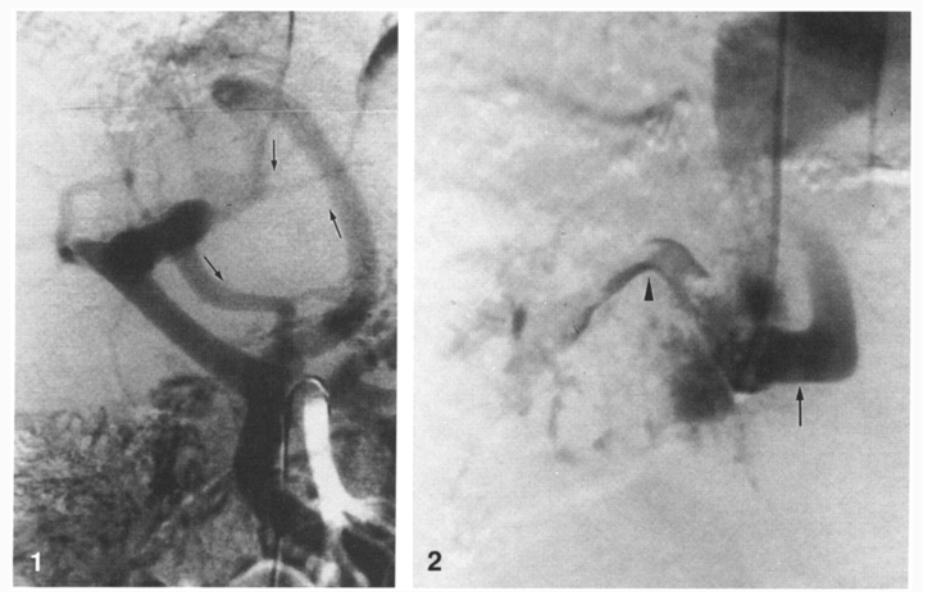

Fig. 1. Superior mesenteric arteriography confirmed a large portosystemic intrahepatic shunt between the left portal and left hepatic vein (arrows).

Fig. 2. Hepatic transjugular phlebography performed by a right jugular approach confirmed occlusion of the right hepatic vein by thrombus (arrowhead) and reflux in the left hepatic vein draining the intrahepatic portosystemic shunt (arrow).

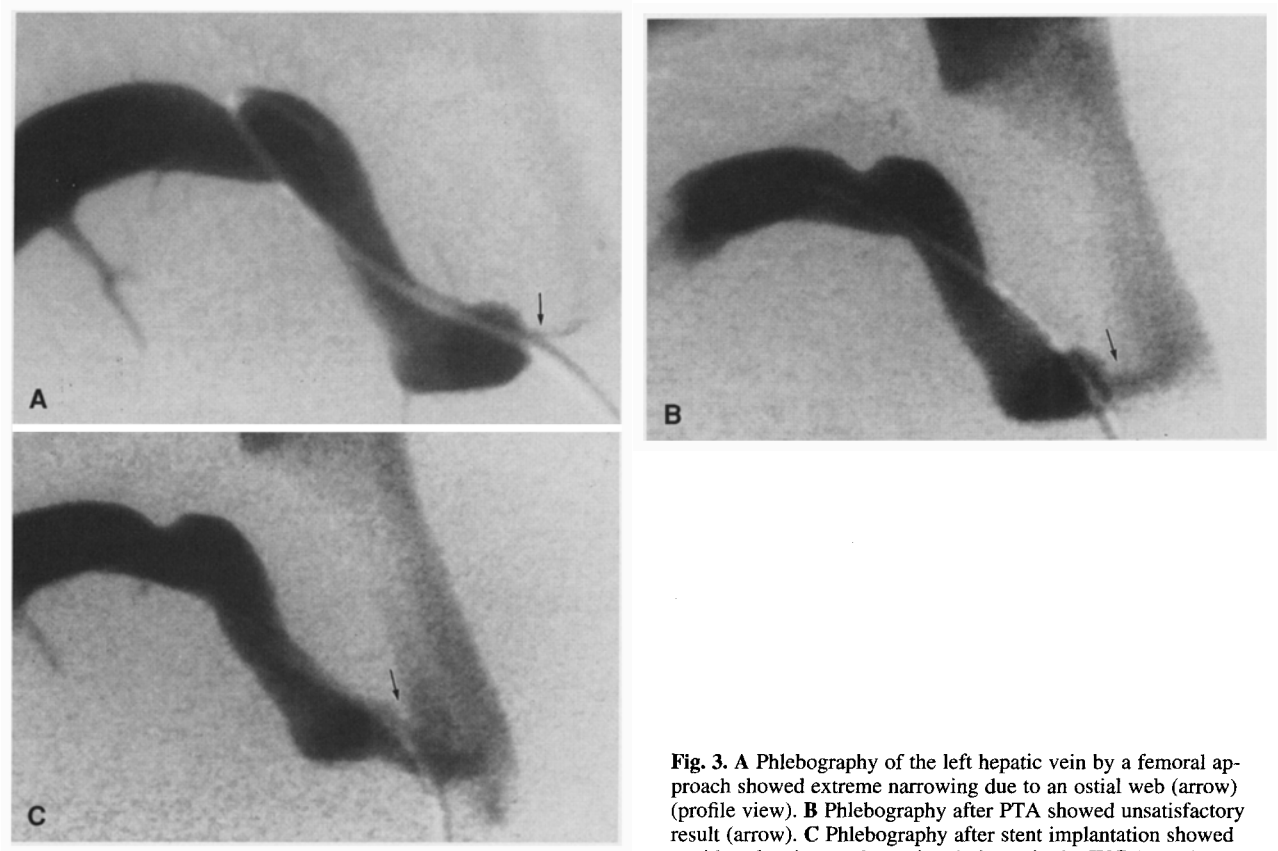

Fig. 3. **A** Phlebography of the left hepatic vein by a femoral approach showed extreme narrowing due to an ostial web (arrow) (profile view). **B** Phlebography after PTA showed unsatisfactory result (arrow). **C** Phlebography after stent implantation showed a widened ostium and massive drainage in the IVC (arrow).

and oral contraceptives, trauma, connectivitis, angiitis, venoocclusive disease caused by alkaloids of senecio, bone marrow transplantation, and anticancer drugs. Stenoses or occlusive membranous webs may be of developmental origin, but preexisting thrombosis can also result in intimal fibrosis, lumen narrowing, and endovascular membranes [1].

Budd-Chiari syndrome caused by diffuse thrombosis and with impairment of hepatic function is treated by liver transplantation or portocaval shunts. Patients with other

Table 1. Budd-Chiari syndrome treated by expandable metal stents

Year [Reference]	Number of cases	Type and distribution of stenosis	Type and number of stents	Technique	Follow-up (months)
1990 [4]	1	LHV: obstruction	1 Palmaz stent	2 months after 3 PTAs	12
1990 [5]	2	IVC: membranous web			12
1990 [6]	3	IVC: thrombosis	1 GS	Immediately after PTA	10
			2 single and 1 double GS	3 months after PTA	10
			3 double GSs	Immediately after PTA	7
1991 [7, 13]	1	IVC: membranous web	1 double GS	Immediately after PTA	44
		LHV: membranous web	1 triple CS	After PTA	44
1991 [8]	1	HV: membranous web	1 Wallstent	Four months after PTA	3
1992 [9]	1	IVC: stenosis and thrombosis	1 GS	Immediately after PTA	21
1990 [10]	1	IVC: stenosis and thrombosis	1 Wallstent	After PTA and thrombolysis	5
1992 [11]	1	IVC: long segmental stenosis and thrombosis	2 double CSs	After thrombolysis and PTA (one session)	14
1993 [12]	3	HV: segmental obstruction	GS	After PTA	6–18
	3	HV: segmental obstruction	GS	After PTA and thrombolysis	6–18
1994 [14]	4	IVC: segmental obstruction	GS	After PTA	30–44 (3 cases)

LHV = left hepatic vein, IVC = inferior vena cava, HV = hepatic vein, GS = Gianturco stent, PTA = percutaneous transluminal angioplasty

causes who do not respond to medical treatment are also referred for surgery. Our patient was scheduled for liver transplantation, considering persistent abnormal liver tests. Membranous obstruction or the long segmental stenosis of the hepatic veins or the retrohepatic IVC can be corrected by interventional radiological procedures. Anatomical distribution of the lesions was classified in patients from the Far East, providing a guideline for therapy [3]. PTA has been reported in 60 patients with stenoses or limited obstruction. Ten percent to 15% of patients with an anatomical stenosis or web also exhibited predisposing factors, including polycythemia vera, lupus, oral contraceptives, postpartum status, and Behçet disease, illustrating the complex pathoetiology of the disease. The mean number of PTAs necessary to maintain venous patency was 2–3 per patient [2]. Multiple balloons inserted by a bifemoral approach or disruption of the web by pulling back the balloon catheter are possible variants of conventional PTA. Excellent short-term results were obtained in most patients, particularly in membranous webs, with a patency of 81% after 2 years. However, when all causes of Budd-Chiari syndrome are considered, overall patency after PTA was only 50% after 6 months. Reocclusion of a long segmental stenosis of the IVC was the rule at one year, despite prolonged anticoagulation and repeated dilatation.

Considering the poor midterm results and cost of repeated PTA, vascular metal stents have been advocated for prevention of restenosis since the initial report by Walker et al. [4]. Reports on 21 patients treated with metal stents have been published in the literature to the best of our knowledge (Table 1) [4–14]. Either the IVC [5, 6, 9, 10, 11, 14] or hepatic veins [4, 8, 12], or both [7] were stented. Intrahepatic venous connections allow decompression of venous outflow by restoring flow in the dominant hepatic vein only [4, 7], as in our patient. No restenosis had occurred after follow-up of 3–44 months, but secondary thrombosis occurred twice at 3 months [12]. It is not clear if patients should be maintained on anticoagulation [7] or urokinase infusion for several days [9] after stent placement. Limited caval narrowing of 3–5 mm was observed after a 21-month follow-up, and did not progress to occlusive stenosis [6, 12]. It has been suggested that intimal thickening is less pronounced and occurs later with an open stent configuration, such as the Gianturco-type Z-stent [6]. Stents to be placed in the distal IVC or hepatic veins should have a diameter of 1.5–2 times the venous diameter. The stent must be long enough to be maintained in the web without displacement [15]. Whatever type of stent is used, long-term effects on hepatic veins or the IVC have to be further investigated. In four patients, the stent was placed immediately after unsuccessful PTA [6, 7, 9]. Percutaneous placement of stents did not add complications to PTA. Local fibrinolytic infusion is an adjunctive therapy in case of long segmental thrombosis above a tight stenosis [10, 11, 12]. Stents may also be placed without previous thrombolysis, when volume and extent of clot formation is limited [9]. Protrusion of stents from the hepatic vein or stents placed in the distal IVC may cause technical difficulties for liver transplantation. The risk-benefit ratio must be taken into consideration for each patient.

Optimal indication for stenting is secondary restenosis or recoil after initial PTA, as observed in our patient, but considering the nonsatisfactory results of PTA, stents may be inserted without delay after unsuccessful PTA. Transjugular intrahepatic portosystemic stent shunting (TIPS) is another adjuvant technique for treating Budd-Chiari syndrome in selected patients

with advanced disease, when no hepatic vein lumen is identifiable beyond the ostium.

References

1. Kage M, Arakawa M, Kojiro M, Okuda K (1992) Histopathology of membranous obstruction of the inferior vena cava in the Budd-Chiari Syndrome. Gastroenterology 102:2081–2090
2. Martin L, Dondelinger RF (1993) The treatment of Budd-Chiari syndrome by percutaneous transluminal angioplasty. Rev Im Med 6:31–36
3. Hirooka M, Kimura C (1970) Membranous obstruction of the hepatic portion of inferior vena cava. Surgical correction and etiological study. Arch Surg 100:656–663
4. Walker HS, Rholl KS, Register TE, Van Breda A (1990) Percutaneous placement of a hepatic vein stent in the treatment of Budd-Chiari Syndrome. J Vasc Intervent Radiol 1:23–27
5. Takahashi M, Okawada T, Keneko M, Xu KI, He-Fang X (1992) Budd-Chiari syndrome of unknown origin studied by international cooperative research programme (Scientific Exhibit). Cardiovascular and Interventional Radiological Society of Europe (CIRSE), Annual Meeting, Sitges, Spain
6. Furui S, Sawada S, Irie T, Makita K, Yamaguchi T, Kusano S, Ibukuro K, Nakamura N, Takenaka E (1990) Hepatic inferior vena cava obstruction: Treatment of two types with Gianturco expandable metallic stents. Radiology 176:665–670
7. Lopez RR Jr, Benner KG, Hall L, Rosch J, Pinson CW (1991) Expandable venous stents for treatment of Budd-Chiari Syndrome. Gastroenterology 100:1435–1441
8. Weernink EE, Huysman AB, Tennappel CHH (1991) Treatment of Budd-Chiari syndrome by insertion of Wallstent in hepatic vein. (letter) Lancet 338:644
9. Sawada S, Fujiwara Y, Koyama T, Kobayashi M, Tanigawa N, Iwamiya T, Katsube Y, Nakamura M, Furui S (1992) Application of expandable metallic stents to the venous system. Acta Radiol 33:156–159
10. Fujihara Y, Sawada S, Koyama T, Tanabe Y, Saitou S, Katsube Y, Saitu M (1990) Clinical experience of percutaneous transluminal angioplasty and expandable metallic stent placement for Budd-Chiari syndrome with massive thrombus in the inferior vena cava: Report of a case. Rinsho-Hoshasen 35:1435–1438
11. Ishiguchi T, Fukatsu H, Itoh S, Shimamoto K, Sakuma S (1992) Budd-Chiari syndrome with long segmental inferior vena cava obstruction: Treatment with thrombolysis, angioplasty and intravascular stents. J Vasc Intervent Radiol 3:421–425
12. Park JH, Chung JW, Choi CK, Han JK, Han MC (1993) Benign obstruction of hepatic inferior vena cava: Imaging, diagnosis and intervention. Radiology, 189 (P):358
13. Rosch J, Keller FS, Uchida BT, Barton RE (1994) Interventional management of large venous obstruction. In: Strandness E, Van Breda A (eds): Vascular diseases. Churchill Livingstone, New York, p 1006
14. Yamada R (1994) Angioplasty of Budd-Chiari syndrome. European Society of Gastro-Intestinal Radiologists, Fifth Annual Meeting, Taormina, Italy, June 1–4
15. Dondelinger RF, Goffette P, Kurdziel JC, Roche A (1991) Expandable metal stents for stenoses of the venae cavae and large veins. Semin Intervent Radiol 8:252–263