Primary Budd-Chiari syndrome: Outcome of endovascular management for suprahepatic venous obstruction

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Objective: **Primary Budd-Chiari syndrome (BCS) is a rare form of hepatic venous outflow obstruction at the suprahepatic inferior vena cava (IVC), the hepatic veins, or both. We assessed our 4-year experience in the management of BCS to evaluate the results of our methods of care.**
Methods: **We conducted a retrospective review of a nonrandomized clinical trial conducted in three teaching hospitals. Among 28 primary BCS patients, 9 remained in medical treatment only, and 19 who failed to respond to medical treatment received additional endovascular (n = 17) or surgical therapy (n = 2). Nine underwent IVC balloon angioplasty alone, 6 had angioplasty plus stents, and 2 had transjugular intrahepatic portosystemic shunts (TIPS) for hepatic vein lesions. One patient had a mesoatrial bypass; another had liver transplantation. Immediate response to the therapy was assessed with angiography and ultrasonography based on anatomic and/or hemodynamic correction or reduction of the lesion. Subsequent assessment of portal hypertension status was made with periodic clinical and laboratory evaluation (eg, ultrasonography, liver biopsy).**
Results: **Twenty-six patients had had IVC stenosis or occlusion by focal or segmental lesion. Two patients had hepatic vein outlet obstruction. There was no evidence of coagulopathy as the pathogenesis; all were related to membranous obstruction of the vena cava. Excellent immediate response to the endovascular therapy and subsequent relief of portal hypertension were achieved in 14 patients. Four patients had restenosis or progression of the residual lesion within 2 years; three responded to repeated stenting. Primary patency was 76.5%, and primary assisted patency was 94.1%. Two patients with TIPS and two with surgical therapy maintained excellent results. The medical treatment remained effective only in a limited group of 6 (21.4%) of the 28 patients.**
Conclusions: **In BCS, both endovascular and surgical interventions provide excellent results and potentially halt liver parenchymal deterioration caused by portal hypertension. Liver transplantation remains the ultimate solution for advanced liver failure. (J Vasc Surg 2006;43:101-8.)**

Hepatic venous outflow obstruction at the suprahepatic inferior vena cava (IVC), the hepatic veins, or both, has been known as Budd-Chiari syndrome (BCS) regardless of its etiologies (eg, primary and secondary) and pathogenesis (eg, congenital, inflammatory, infectious origin, or a combination).[1-6] Primary BCS is a relatively rare condition, with geographic variances in etiologic and predisposing factors, with striking difference between African and Asian patients and Caucasian (Western) patients.[4-9] The developmental anomaly theory is mostly compatible with African and Asian patients as its etiopathogenesis, while the thrombosis theory is more compatible to Caucasian patients.[4-7]

Management strategy also remains critical and includes decompression procedures vs curative radical procedures.[10-13] The optimum combination of various surgical and endovascular procedures[14-17] is also not quite clear, especially when BCS is considered as a precancerous lesion.[18-20]

We assessed our experiences in the management of suprahepatic IVC, hepatic vein obstruction, or both, by primary BCS to evaluate the results of our methods of care.

METHODS AND PATIENTS

A retrospective analysis of 28 patients (13 men, 15 women; mean age, 43.2 years) was conducted for a 4-year period between September 1994 and August 2001 at Samsung Medical Center and its affiliated hospitals of Sungkyunkwan University, Seoul, Korea. Nineteen patients (11 men, 8 women; aged 28 to 68 years; mean, 45 years) who failed to respond to medical therapy with evidence of progress for a minimum 1-year period were selected for endovascular, or surgical treatment, or both. The other nine patients (2 men, 7 women; aged 24 to 59 years; mean, 41.4 years) with a satisfactory response to the medical therapy remained in the medical therapy group.

Diagnostic evaluation of BCS included functional and anatomic assessment of the IVC and hepatic veins with laboratory and chemical tests as well as other noninvasive tests. In addition to liver chemistry and hepatitis profile, a

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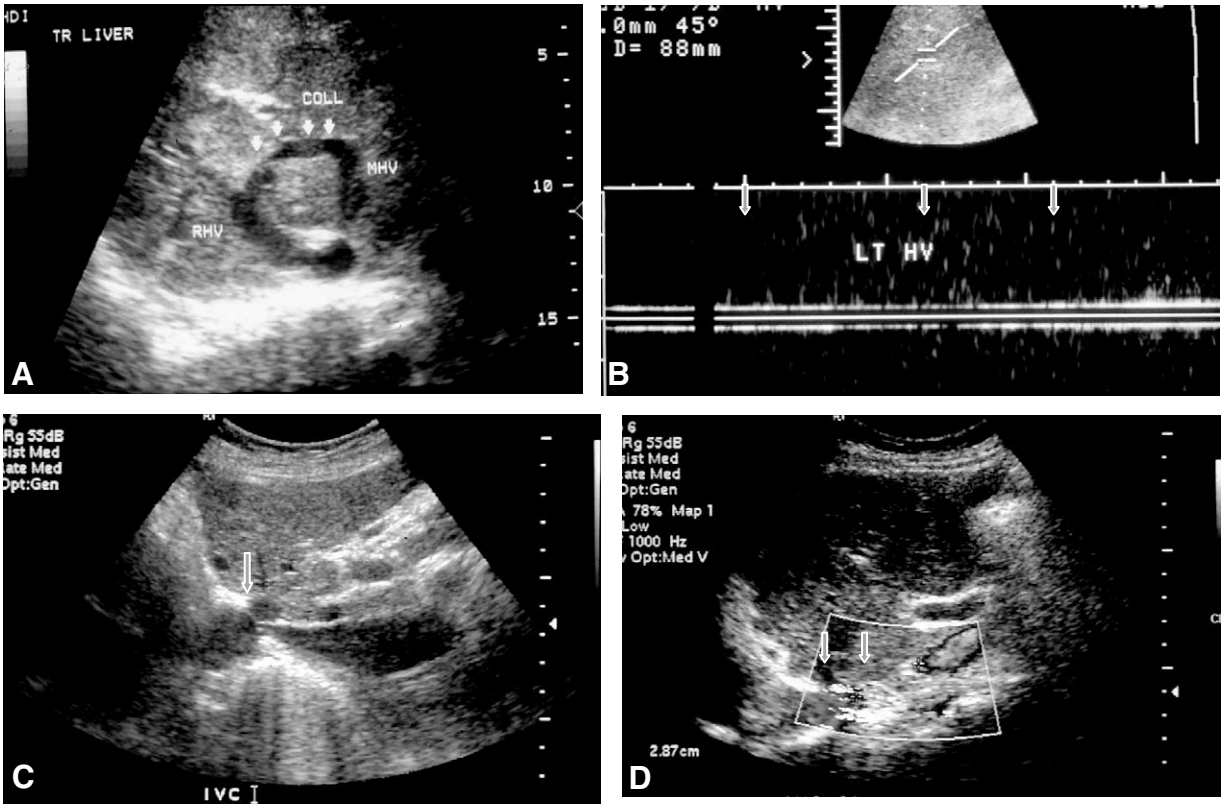

Fig 1. Duplex ultrasonographic findings of various intrahepatic conditions due to primary Budd-Chiari syndrome (BCS). A, Collateral vein is displayed as typical "ring" shape connection *(white arrows)* between right *(RMV)* and middle hepatic veins *(MHV)*, reflecting hepatic venous outflow obstruction. B, Left hepatic vein demonstrates a lack of blood flow following the thrombosis. C, Focal stenosis of suprahepatic inferior vena cava *(IVC)* is shown as the cause of hepatic venous outflow obstruction. It is most probably due to the membranous obstruction of the IVC, the most common cause of primary BCS. D, Segmental stenosis of IVC (2.87 cm) is shown with evidence of portal hypertension.

coagulation profile was assessed in all patients to rule out the coagulopathy as predisposing factor.

Duplex ultrasonographic studies[21-23] and contrast-enhanced computed tomography (CT) scans [24] of IVC and hepatic veins were performed in all 28 patients to assess the status of the intrahepatic venous collateral and portal vein system and hepatic venous outflow along the IVC. Signs of obstruction of hepatic venous outflow, abnormalities in the direction of flow in the hepatic veins, and communicating vessels between the hepatic veins were assessed with real-time imaging by the pulsed Doppler analysis (Fig 1). Magnetic resonance (MR) imaging or MR angiography,[25] or both, a were added selectively when indicated.

A baseline liver biopsy specimen was obtained in most patients, but a follow-up biopsy was performed only when evidence showed that the disease was progressing. Transfemoral IVC angiography was performed in all patients as a road map for the further treatment procedure.

Selective portal vein studies with direct splenoportography, celiac-hepatic angiography, percutaneous transhepatic hepatic venography, or a combination, were limited to those candidates for a transjugular intrahepatic portosystemic shunt (TIPS) or other surgical decompression procedures for portal hypertension.[10,11]

The medical therapy included general care for portal hypertension, liver cirrhosis, and ascites with diuretics, hepatotonic measurement with nutritional support, and empirical treatment of various symptoms and signs of portal hypertension. Anticoagulation therapy was not included. The response to medical treatment was assessed every 6 months with biochemical liver function tests and ultrasonographic evaluation of liver parenchymal status.

Among 19 patients, 17 received endovascular treatment and dilatation to the suprahepatic IVC or hepatic vein lesions, or both. Two underwent surgery—bypass for the complete obstruction of long segment of suprahepatic IVC with massive thrombosis, and transplantation for advanced cirrhosis—because endovascular treatment was neither indicated nor technically feasible.

Nine patients had IVC balloon angioplasty alone to the focal (n = 8) or short (<2.5 cm long) segmental (n = 1) stenosis caused by membranous obstruction lesions of the IVC (MOVC). Six who did not adequately respond to the initial angioplasty had simultaneous stent procedures: two

had focal and four had segmental stenosis of the IVC.[14-17] Two had TIPS to relieve hepatic venous outlet obstruction caused by lesions between IVC and hepatic veins.[11]

Inferior vena cava balloon angioplasty was done mostly by the transfemoral approach, except in the patients who required the "kissing balloon technique" via a simultaneous transjugular approach. Various angioplasty catheters (5- to 18-mm balloon) (Cook, Bloomington, Ind) were used for the preliminary and final dilatation together with conventional spring-coil and laminated hydrophilic guide wires with floppy tips or stiff end. Wallstents (Boston Scientific, Natick, Mass) were used for the stenting procedures.

One patient, who had a history of failed angioplasty and occlusion of IVC with extensive thrombosis, had a mesoatrial bypass[26,27] from the superior mesenteric vein to the right atrium. An 18-mm ringed polytetrafluoroethylene graft was used to relieve progressive liver cirrhosis and portal hypertension. Another patient, who had a failed/thrombosed cavoatrial bypass done 2 years before the referral, had cadaver donor liver transplantation[10,12]for a rapidly deteriorating liver condition.

The immediate results of the treatment were assessed for anatomic and hemodynamic responses to the procedures:

- Excellent—anatomically complete to near-complete dilatation of the stenosis and/or hemodynamically comparable reduction of the pressure gradient;
- Good—substantial correction of stenosis with minimal to 25% to 30% residual lesion anatomically and/or comparable residual pressure gradient;
- Fair—significant (<50%) residual lesion with improved pressure gradient;
- Poor—substantial (>50%) residual lesion with minimum to no pressure gradient improvement.

For the endovascular procedures, heparin was used only during the procedure unless simultaneous thrombolysis was required; heparin use was then extended for 1 week, followed by sodium warfarin for 1 month (international normalized ratio, 2.0). All other routine endovascular patients were on the antiplatelet agent for only 1 month after the procedures. The patient who had bypass surgery was also prescribed an antiplatelet agent for 1 month.

Follow-up assessment for the treatment results was made of all 19 patients, with a clinical evaluation as well as an ultrasonography study and liver function tests, starting at 2 weeks and continuing at 1 month, 2 months, and 3 months, and then at 6-month intervals thereafter for a minimum of 4 years. Additional tests (eg, CT/MR angiography, liver biopsy) were performed when indicated.

Clinical criteria for the relief of portal hypertension following the treatment include:

- Excellent—complete to near-complete relieve of ascites and/or hepatomegaly with compatible laboratory findings (eg, disappearance of hepatic parenchymal congestion on ultrasound evaluation);
- Good—substantial relief of clinical evidence of portal hypertension with compatible laboratory improvement; and
- Fair—noticeable responses with minimal clinical improvement.

Liver parenchymal status was separately assessed by duplex ultrasound study in all 19 patients. Parenchymal congestion was graded as *remained*, *improved*, *relieved*, or *progressive* by two independent radiologists. A liver biopsy, mostly done percutaneously, was also included in 10 patients with known cirrhosis to evaluate the treatment response. Cirrhosis was graded as *improved*, *stable*, or *progressive* by the histopathologists.

RESULTS

All 28 patients were confirmed as having the primary type of BCS due to suprahepatic IVC stenosis or occlusion in 26 patients or hepatic vein outlet obstructive lesions in two patients on ultrasonographic study. All the IVC lesions were either focal stenoses or occlusions with or without membranous lesions in 14, or segmental stenoses or occlusions in 12. All IVC lesions had at least one patent hepatic vein opening, mostly with the right hepatic vein in 22 into the IVC below the site of occlusion; the right hepatic vein had generally extensive collateral channels of intrahepatic veins to communicate with left or middle hepatic vein (Fig 1).

Other etiologic and predisposing factors, especially hypercoagulopathy, were completely ruled out in the 28 patients. They had an average of 3.6 years (2.8 to 4.1 years) of clinical history reflecting chronic portal hypertension before the diagnosis of primary BCS, but their liver profiles showed remarkably well-preserved liver function, except for moderate to severe hepatic congestion.

They have shown various clinical symptoms and signs; ascites was the most common finding together with hepatomegaly (n = 24, 86%), back or flank collateral veins was the second most common findings (n = 20, 71%), pain or ache in the abdomen (n = 18, 64%), splenomegaly (n = 14, 50%), jaundice (n = 9, 32%), and gastrointestinal bleeding (n = 2, 7%). Most were remarkably free from chronic venous hypertension of the lower extremity, only a small group of six patients had minimal leg swelling, and there were no cases of venous claudication.

Procedures in the 17 endovascular patients were technically successful, with excellent to good responses as immediate outcomes and subsequent relief of portal hypertension in 14 (82%) (Fig 1): angioplasty alone (78%) to the focal stenosis in 7, angioplasty plus stent (83%) to the focal and segmental stenosis in 5, and TIPS (100%) to the hepatic vein outlet obstruction in 2.

Nine patients were treated with angioplasty alone, and all eight focal lesions showed immediate or technical success with excellent results in seven and good results in one. One segmental lesion had fair response, but the patient refused to have additional stenting simultaneously to improve the result.

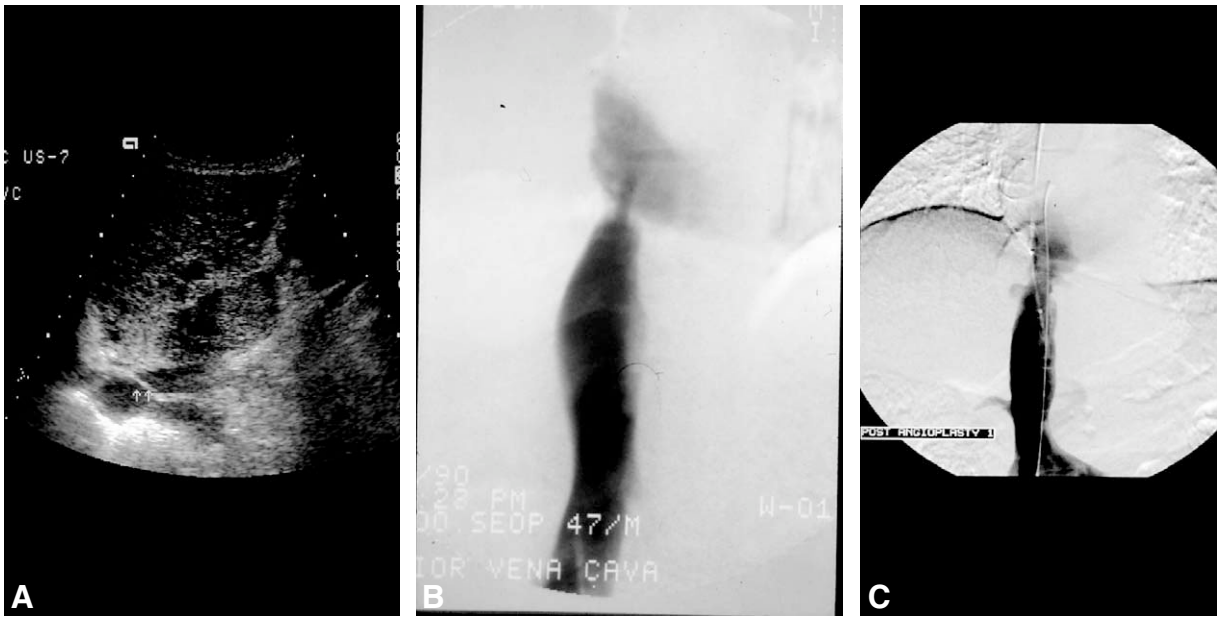

Fig 2. Endovascular management of focal stenotic lesion with balloon angioplasty alone. A, Duplex scan demonstrates membranes *(arrows)* as the cause of obstruction; it depicts focal stenosis of the inferior vena cava (IVC) as the cause of Budd-Chiari syndrome. B, Angiographic finding displays a tight stenosis of suprahepatic IVC. C, Shows the excellent hemodynamic response to angioplasty alone to relieve portal hypertension subsequently; a residual stenosis of 31% remains despite complete reduction of the pressure gradient.

In the six patients with angioplasty plus stent, excellent immediate or technical success was achieved in the two focal lesions, and three of four segmental lesions also had excellent responses. The response in on segmental lesion remained poor to minimum, but the patient refused to have a retrial of the stent procedure (Fig 2). The average pressure gradient of 28.2 mm Hg was reduced to 10.6 mm Hg by the successful procedures.

Two patients were treated with TIPS using 10-mm diameter Wallstents; both had excellent responses, with immediate relief of hepatic vein outlet obstruction. The pressure gradient has shown a substantial reduction, from 29.1mm Hg to 10.2 mm Hg (patient 1) and 23.8 mm Hg to 9.2 mm Hg (patient 2) (Fig 3).

All 14 patients (angioplasty alone in 7, angioplasty plus stent in 5, and TIPS in 2) who had excellent technical response to the endovascular procedures subsequently achieved excellent relief of portal hypertension within 6 months, with compatible improvement of clinical (eg, ascites and hepatomegaly) and laboratory findings (eg, liver profiles, hepatic congestion finding on ultrasound study). Among three other patients, two showed fair to minimum improvement by clinical and laboratory evaluations at the subsequent 6-month follow-up, and one, with advanced cirrhosis, showed poor to no response, which were compatible with ultrasound findings.

In four patients in the endovascular group (n = 7), restenoses or progression of residual stenosis developed within 2 years along the region previously treated with endovascular dilatation, for a failure rate of 23.5%.

Among the patients treated with angioplasty alone, one with a segmental lesion who had a fair response to the inial treatment showed deterioration within 6 months and later underwent an additional stenting procedure; another with a focal lesion that responded well to the initial angioplasty was also found to be in near-occlusive status after she missed her regular follow-up assessments for 1 year. Two patients treated with angioplasty plus stent for the segmental lesions, including one who failed to respond initially, were found to have further progress of the lesion after they missed follow-up for 2 years. However, all four patients immediately underwent reintervention for the occlusion or restenosis of IVC with a stent, two of which required combined thrombolytic therapy. Three patients had an excellent outcome, but one did not improve after this second endovascular procedure and refused bypass surgery despite steady deterioration. Therefore, during a minimum 4-year follow-up (average, 4.5 years), 17 endovascular procedures had a primary patency rate of 76.5%, and the primary assisted patency rate was 94.1%. No deaths occurred among this endovascular group during the follow-up period.

Mesoatrial bypass achieved immediate reduction of portal pressure from 50 to 8 cm H_2O intraoperatively and maintained excellent patency, with complete relief of portal hypertension (Fig 4). Steady clinical improvement, such as disappearance of ascites, abdominal wall collaterals, and hepatomegaly, was accompanied by compatible improvement of the liver status as confirmed by periodic ultrasound studies and liver biopsy specimens (4.6 years).

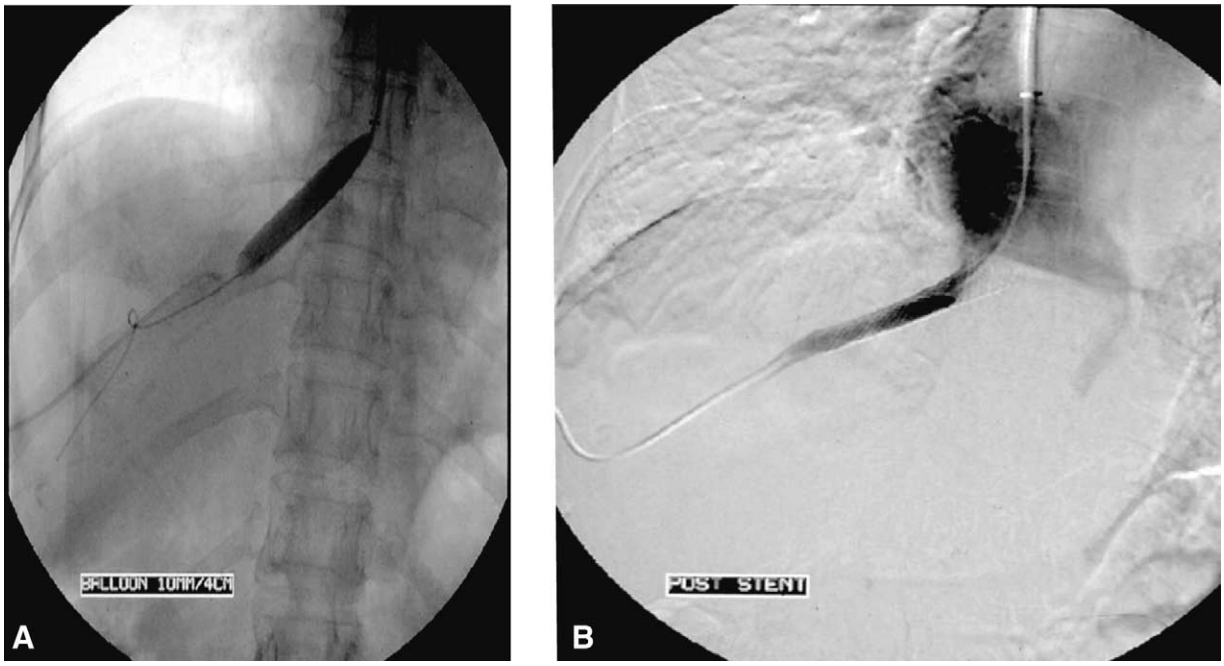

Fig 3. Endovascular management of hepatic vein outlet obstruction with angioplasty plus stent procedure. A, Angiographic finding of balloon angioplasty from the hepatic vein outlet to the inferior vena cava (IVC), which is the initial procedure to place a transjugular intrahepatic portosystemic shunt (TIPS) to relieve portal hypertension. B, Angiography depicts successful stenting procedure over the balloon-dilated and newly established portosystemic shunt.

The liver transplant patient also maintained excellent function throughout the follow-up period (4.2 years), and the transplanted liver had no evidence of BCS or cancer development. The recipient's explanted liver was found to have early stage hepatocellular carcinoma (HCC).[18-20]

Among the endovascular/surgical group, ultrasound evaluation of liver parenchymal congestion showed 15 relieved, 3 improved, and 1 progressed, which were directly proportional to the anatomic and hemodynamic response to the therapy.

The liver biopsy specimens from 10 cirrhotic patients showed a variable degree of outflow obstruction and hepatocellular damage from sinusoidal dilation with centrilobular congestion to hepatocyte necrosis. But the findings of mild to moderate fibrosis were common in the majority. Among these 10 patients, 5 remained histologically stable, including 1 who had an unsatisfactory response to the angioplasty alone, and 2 who improved[28] after endovascular/bypass therapy. Three patients deteriorated, including one with good immediate response and subsequent successful decompression of portal hypertension. None of these three showed evidence of coinfection with hepatitis B and C viruses. Two developed de novo HCC during the follow-up period of 4.7 years and are currently on chemoembolization therapy.

Nine patients remained in medical treatment, of which six were able to maintain a stable liver condition during the 4.9-year follow-up period. However, three have shown progressive deterioration of liver condition, documented by the liver biopsy and ultrasound study, despite adequate medical treatment; all three refused additional surgical or endovascular treatment. In one of these three patients, HCC was reported to have developed soon after the patient stopped the follow-up assessment (4.7 years), and the patient was reported to have died 1 year after the diagnosis. Therefore, the overall response to the medical treatment only during the limited follow-up period was 21.4% (6/28) (Table).

DISCUSSION

Budd-Chiari syndrome is a generic term for various forms of portal hypertension secondary to obstruction of hepatic venous outflow, with or without combined IVC obstruction. Although MOVC [5,7] is known as the most frequent cause of primary BCS among Africans and Asians, the hypercoagulable state[4,9] remains the most important cause for Caucasians. Full screening for hypercoagulability, including collagenous vascular disease, is therefore essential for the investigation of all the primary BCS. Careful assessment of all other possible causes, such as bacterial infection, and Behçet's disease,[29,30] also needs to be included to determine any predisposing factors.

Most of our patients had MOVC-related IVC, hepatic vein obstruction or stenoses, or both, similar to other reports from Asia.[7,14,17] This unique condition among our patients supports the congenital/developmental anomaly theory of primary BCS for its etiopathogenesis.[4,5,7] There have been many reports on the con-

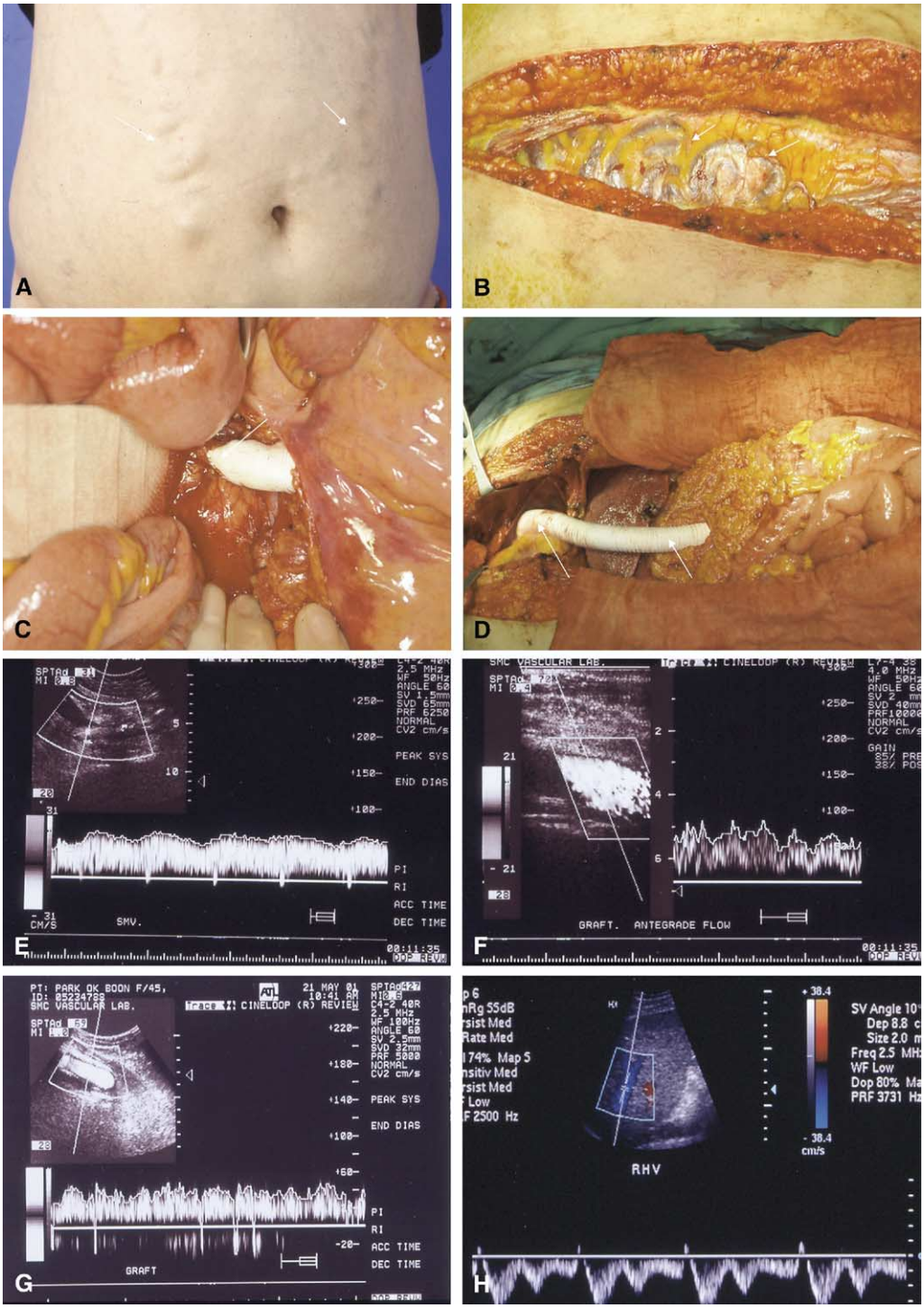

Fig 4. Surgical management of inferior vena cava (IVC) occlusion with mesoatrial bypass to relieve portal hypertension. A, Clinical appearance depicts extensive collateral veins *(arrows)* to compensate IVC occlusion along the distended abdominal wall with ascites. B, Operative finding shows extensive collateral veins *(arrows)* through the properitoneal layer of abdominal wall. C, End-to-side anastomosis of polytetrafluoroethylene ring graft *(arrow)* to the superior mesenteric vein is displayed as a part of mesoatrial bypass surgery to decompress portal hypertension. D, Mesoatrial bypass graft *(arrows)* is being led into the mediastinal space anteriorly to the liver for the proximal anastomosis to the right atrium. E, Duplex ultrasonographic evaluation depicts excellent blood flow through the superior mesenteric vein after successful decompression of portal hypertension with bypass. F, Excellent antegrade blood flow through the graft demonstrates well-functioning graft (G) diverting the portal blood flow to the right atrium. H, Fully restored blood flow within the right hepatic vein displays successful decompression of portal hypertension by the bypass.

Clinical summary of primary Budd-Chiari syndrome management outcome in 28 patients.

1. Medical treatment group (n = 9); follow-up, 4.9 years
   - n = 6: maintained clinically stable condition with comparable liver status.
   - n = 3: deteriorated progressively but refused to receive additional therapy.
   - n = 1: reported to have developed liver cancer later.
2. Endovascular treatment group (n = 17); follow-up, 4.5 years
   A. Balloon angioplasty alone (n = 9)
   - n = 8: excellent to good immediate response and subsequent relief of portal hypertension.
   - n = 1: fair response to the first procedure with angioplasty alone. Later developed liver cancer despite excellent decompression with subsequent stenting.
   - n = 2: additional stenting to recurred/progressing lesions with excellent results.

   B. Balloon angioplasty plus stent (n = 6)
   - n = 5: excellent immediate response and subsequent relief of portal hypertension.
   - n = 1: poor response with steady deterioration, and failed second stenting. Later developed liver cancer.
   - n = 2: additional stenting to recurred/progressive lesions- 1 failed.

   C. Transjugular intrahepatic portosystemic shunt (n = 2)
   - Excellent response with immediate relief of portal hypertension and subsequent clinical improvement.
3. Surgical treatment (n = 2)
   - n = 1: Mesoatrial bypass; maintains excellent outcome with full decompression of portal hypertension; follow-up, 4.6 years.
   - n = 1: Liver transplantation; maintains excellent outcome with no evidence of Budd-Chiari syndrome recurrence; follow up, 4.2 years.

genital anomalies of IVC, including its length, location, duplication, abnormal connection and draining, and residual remnant of embryonal tissue-like webs[31-37] Especially for the pathogenesis of MOVC, this congenital anomaly theory gains significant support on the basis of such intriguing embryogenesis of IVC with naturally increased risk of developmental anomaly.

Angiography has been the mainstay for diagnosis of BCS, but real-time ultrasonography [21, 22] has shown excellent ability to deliver unique intrahepatic hemodynamic and pathophysiologic status (Fig 1). We were able to identify the abnormalities of right, middle, and left hepatic vein in most patients, in addition to abnormal flow patterns. Uniphasic flow was the most common abnormality, and most had an abnormal waveform in one or more hepatic vein. Intrahepatic collaterals and hepatopetal flow in the portal vein were common findings.

Direct surgical and endovascular intervention seem to provide acceptable palliation for most primary BCS lesions of African/Asian type; this modality of therapy might not be able to provide same results to Caucasian (Western type) primary BCS.. Endovascular intervention was accepted as a safe, less invasive, cost-effective method with less complication in our institute.

If venous outflow cannot be restored interventionally or surgically, and liver function continues to deteriorate or cirrhosis develops, liver transplantation should be considered.

Our experiences with HCC development in 4 (14.3%) of 28 patients with primary BCS is compatible with others reporting a high incidence among African and Asian patients and shows the striking differences among races.[19, 20]

In view of distinctive nature of primary BCS as a precancerous lesion among African/Asian patients with a high incidence of HCC,[18,19] liver transplantation has a unique role in advanced hepatic failure or a steadily deteriorating liver condition caused by the primary BCS. An aggressive approach with liver transplantation may be warranted as a preemptive measure, especially when all the available methods fail to stop the progression of the disease. The judgement solely on the basis of clinical response may be inappropriate, and vigilant surveillance with appropriate screening of HCC during follow-up using an imaging modality such as ultrasound is mandated, especially in African/Asian patients with MOVC as a high-risk group for HCC development.

CONCLUSIONS

In primary Budd-Chiari syndrome, both endovascular and surgical interventions provide excellent results and potentially halt liver parenchymal deterioration caused by portal hypertension. Both surgical and endovascular procedures may prevent hepatocellular carcinoma as a complication of primary BCS. However, long-term follow-up studies are necessary to support this hypothesis.

Liver transplantation would remain ultimate solution for far advanced liver failure.

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Analysis and interpretation: BBL, LV
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Final approval of the article: BBL, LV, YWK, YSD, KCK, HKL, JHL, KWA
Statistical analysis: BBL, LV
Overall responsibility: BBL

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