Radical surgical treatment of Budd-Chiari syndrome through entire exposure of hepatic inferior vena cava

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ABSTRACT

Objective: Therapies for Budd-Chiari syndrome (BCS) can be divided into three main categories: medical, endovascular, and surgical. Surgery is applicable to the disease when other therapeutic options have failed. We introduce a surgical method of recanalization through exposure of the entire hepatic inferior vena cava (IVC) and hepatic vein (HV) outflow tract for BCS and investigate the long-term outcomes.

Methods: From July 2002 to December 2015 in our center, 83 consecutive symptomatic BCS patients with failure of endovascular therapy were treated by radical surgical recanalization. IVC recanalization was the first goal for all patients, and recanalization of at least one HV was the second goal for selected patients at the same surgical operation. Patients were followed up, and data on technical and clinical success, survival, and patency of target vessels were analyzed.

Results: Technical success of surgical recanalization was achieved in 80 patients (96.4%), with relief of clinical symptoms and improvement of liver function. During a mean follow-up of 84 ± 25.9 months, the cumulative 1-, 3-, and 5-year primary patency rates of the HV were 96.7%, 90.0%, and 83.3%, respectively. The cumulative 1-, 3-, and 5-year primary patency of the IVC was 86.7%, 71.7%, and 68.3%, respectively. No factor demonstrated significant association with recurrence of obstruction. During follow-up, 10 patients died, 8 of end-stage hepatic disease and 2 of unknown causes. The cumulative 1-, 3-, and 5-year all-cause survival rates were 91%, 90%, and 87%, respectively. Female sex, encephalopathy, severe ascites, and hypersplenism had an impact on survival in univariate analysis. With Cox regression, encephalopathy was the only independent determining factor for surgical survival.

Conclusions: Surgical recanalization through exposure of the entire hepatic IVC for BCS is suitable for most primary BCS patients after failure of endovascular therapies. (J Vasc Surg: Venous and Lym Dis 2019;7:74-81.)

Keywords: Budd-Chiari syndrome; Hepatic veins; Vena cava, inferior; Surgery

Budd-Chiari syndrome (BCS) is a rare disease caused by complete or partial obstruction of the hepatic vein (HV) outflow at various parts from small HVs to the atriocaval junction. The disease has been found to impair liver function, which is manifested as abdominal discomfort, hepatomegaly, and ascites, and mainly affects young adults, weakening their capacity for physical labor.[1,2] It has been described to occur on average in 1/100,000 of the population worldwide.[3] However, the incidence of BCS in East Asia is higher than in other regions. By the end of 2013, there were >20,191 BCS cases reported in China, with the estimated incidence between 0.88/million and 7.79/million per year in the Yellow River and whole Huai River basin,[4,5] suggesting that the disease has become a threat to local public health.

Classification of BCS is important for treatment choice. According to the different locations of the obstruction, BCS can be classified as three types: inferior vena cava (IVC) type, HV type, and combined type.[6,7] The IVC or combined type is common in Asian countries, whereas the HV type is frequent in Western countries.[1,8,9] At present, endovascular intervention or absolute medical therapy is usually effective for patients with IVC type. However, the therapeutic strategy for HV type or combined type is relatively complex.[1,2,10] Anticoagulant therapy is usually of little value for portal hypertension and complications of liver disease.[11] The primary patency and secondary patency of percutaneous transhepatic balloon angioplasty for BCS secondary to HV outflow were only 76% and 84% at 2 years, respectively,[12] whereas the 2-year primary patency of transjugular intrahepatic portosystemic shunt (TIPS) was 67%.[13] If initial endovascular recanalization and anticoagulation fail, the surgical option should be actively considered to avoid persistent portal hypertension and secondary liver failure. Considering that a wide variety of complications may be encountered, TIPS and liver transplantation are usually indicated as the last methods for the prevention of irreversible liver failure.[1-3,10,14]

Conventional surgical methods include membrane resection, IVC reconstruction with or without a patch,

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and portosystemic shunts. However, portosystemic shunts (portacaval or mesocaval shunts) placed below the diaphragm could fail if the IVC is thrombosed or severely externally compressed, and the application is currently more limited.[1-3,10,14-16] We developed our radical surgical procedure to reconstruct the IVC and HV with resection of the lesions through exposure of the entire hepatic IVC by venovenous bypass with extracorporeal circulation (Supplementary Video, online only).[17] The purpose of this article was to present a >5-year follow-up study on patency and survival in 83 Chinese patients with BCS who received the radical surgery.

METHODS

Each patient received details of operative recanalization and provided written informed consent for the operation. The retrospective study was approved by the ethics committee of Peking University People's Hospital, and further informed consent was waived. Patients' data were identified from electronic databases, electronic medical records, and clinical notes.

Patients. This is a retrospective single-center study using prospectively collected data of consecutive symptomatic BCS patients with interventional therapy failure from July 2002 to December 2015. The definition of symptomatic BCS referred to patients with at least one of the following complications: esophageal and gastric varices; hypersplenism; refractory ascites; and chronic hepatic dysfunction. Patients were excluded if they had BCS secondary to malignant tumor; if they had asymptomatic BCS due to well-established intrahepatic and extrahepatic collateral vessels; or if they underwent TIPS, surgical shunt, or liver transplantation. Baseline characteristics of patients before treatment included age, sex, symptoms, comorbidities, imaging type, disease, and laboratory values.

Diagnostic studies. An extensive hematologic workup was performed that included the usual measurements of blood coagulation, blood cell counts, and hepatic function to evaluate the Child-Pugh class. All patients had Doppler duplex ultrasound and computed tomography of the abdomen. The diagnosis of BCS was based on angiographic demonstration of occlusion of the HVs or IVC. In addition, pressure was measured in the IVC and the right atrium by catheter using a saline manometer positioned at the level of the IVC obstruction during angiography.

Operative procedure. All patients underwent a thoracotomy through the right seventh intercostal space in a left lateral decubitus position. After the pericardial sac was opened, the intrapericardial IVC was exposed. Next, the diaphragm was incised, near the bare area of the liver, toward the IVC, and the entire IVC of the hepatic segment was then carefully dissected. Finally, about 20 cm of the IVC was exposed. After systemic heparinization, a longitudinal cavotomy was then made according to the specific location and nature of the lesion; this may be a membranous lesion, a fresh or organized thrombus, or even a tumor. An incision of 5 cm was usually enough for a membranous lesion with or without fresh thrombus. However, for patients with a long segment of organized thrombus, it was necessary to extend the incision distally. For extensive occlusions of the HVs, a piece of liver tissue covering the outlet of the HVs was sometimes excised for the IVC for HV outlet plasty. A Foley catheter was put into the distal IVC through the incision as a balloon to control the blood flow. Furthermore, a cardiopulmonary bypass machine was used for intraoperative direct blood collection and reinfusion, by which major bleeding from the HVs was recycled from vacuum aspiration and immediately pumped into the body circulation by catheterization of the right atrium. Lesions were completely resected under direct observation. In patients with an IVC constriction, a patch of autologous pericardium was needed to prevent restenosis (Fig 1).

ARTICLE HIGHLIGHTS

- **Type of Research:** Single-center retrospective cohort study
- **Take Home Message:** Technical success of surgical treatment in 83 patients with Budd-Chiari syndrome after failed endovascular therapy was 96%, with 3-year primary patency of the hepatic veins in 90% and of the inferior vena cava in 72%.
- **Recommendation:** Open surgical reconstruction for Budd-Chiari syndrome with exposure of the retrohepatic inferior vena cava is suggested if endovascular therapies fail.

Follow-up. All patients should receive anticoagulation after surgery with warfarin for at least 12 months. Patients were observed in the clinic every 3 months for the first postoperative year and every year thereafter. At each visit, clinical status was determined and blood samples were collected for necessary laboratory tests. Patency of the HV and of the IVC to the atrium was determined every year by Doppler duplex ultrasound, venous angiography with pressure measurements, or both.

Statistical analysis. Continuous variables are summarized as mean ± standard deviation or median. The paired-samples *t*-test or Wilcoxon test was performed to compare variables before and after treatment. Categorical variables were compared by χ^2 test or Fisher exact test. Cumulative patency was calculated using Kaplan-Meier curves. A P value $<.05$ was considered statistically significant. All statistical calculations were performed with SPSS 18.0 (SPSS Inc, Chicago, Ill).

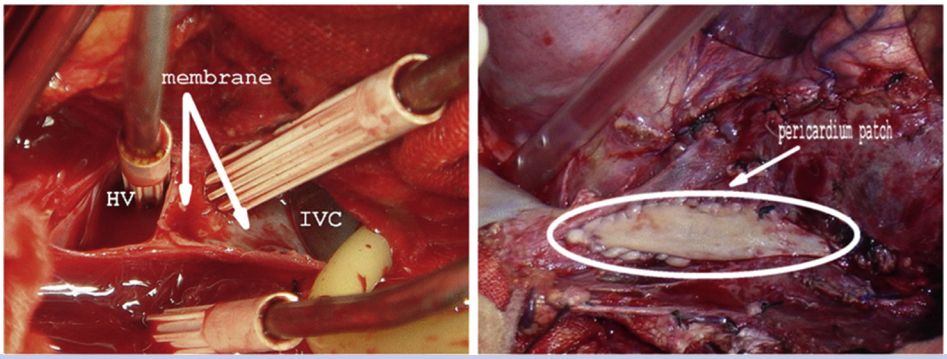

Fig 1. *Left*, Photograph of the operative view with entire exposure of the hepatic inferior vena cava (*IVC*). *Right*, Plasty of IVC for hepatic vein (*HV*) outlet with pericardial patch to prevent restenosis.

RESULTS

A total of 83 patients with BCS were enrolled in this study. The baseline data of patients are demonstrated in the Table. The mean age was 40 ± 12.4 years (range, 13-72 years). Male patients accounted for about 66.3%. Most patients presented with symptoms of portal hypertension, such as ascites (44.6%), hypersplenism (67.5%), jaundice (59.0%), hepatomegaly (71.1%), or superficial epigastric vein varicosis (51.8%). The median course of BCS diagnosis is about 17 months (range, 0.5-384 months). Obstruction was the HV type in 30.1% (25/83) of patients, the IVC type in 15.7% (13/83), and the combined type in 54.2% (45/83). The pathologic process of BCS in the 83 patients included thrombus (13/83 [15.87%]), membrane (22/83 [26.5%]), and combined component (48/83 [57.8%]). The mean follow-up was 84.1 ± 25.9 months, and four patients were lost.

Technical and clinical success and procedure-related complications. Technical success of surgical recanalization of the IVC or HV was achieved in 80 patients (96.4%). Three technical failures were combined-type obstruction cases; two patients had long segmental obstruction of the IVC, and angiorrhaphy rather than recanalization was applied because of intraoperative severe IVC rupture and hemorrhage. Another patient with comorbid Behçet disease suffered a severe liver laceration during operation and received hepatorrhaphy but died 12 hours after surgery.

Except for these three patients, relief of clinical symptoms and liver function improvement after technical success were achieved in all 80 patients. During the early 30 days after operation, three patients presented with symptomatic relapse. The clinically recurrent cases included one with HV obstruction and two with combined obstruction. The patient with HV-type clinical failure died of hematemesis caused by variceal rupture 14 months after the procedure. The other two patients with combined obstruction died 7 months and 11 months after the procedures, respectively, as a result of hepatic failure. The total clinical success rate was 92.8% (77/83).

Three other patients encountered procedure-related complications. One patient with extensive HV obstruction had pulmonary embolism 24 hours postoperatively, and symptoms were relieved after catheter thrombolysis. Another two patients with combined obstruction developed acute cardiac failure, and the reason might be the increase of venous return after recanalization.

Survival. Five patients died during follow-up. BCS-related deaths were due to recurrence of gastrointestinal bleeding (n = 1) and hepatic failure (n = 2). The other causes of death were hepatocellular carcinoma (n = 1) and renal failure (n = 1). The cumulative 1-, 3-, and 5-year survival rates were 94.9%, 89.7%, and 88.4%, respectively (Fig 2).

Factors that influenced survival were assessed with univariate analysis. Child-Pugh class (P = .001), age (P = .046), encephalopathy (P = .001), ascites (P = .003), diabetes (P = .052), and splenomegaly (P = .046) had an impact on outcome. However, on multivariate analysis, the factors chiefly selected were significant on the univariate analysis; Child-Pugh C score was the only independent determining factor for surgical survival. Patients with Child-Pugh C score had a significantly higher mortality than those with Child-Pugh A or B score (P = .001; Fig 2).

Patency. A total of 16 patients, including 4 cases of HV type, 4 cases of IVC type, and 8 cases of combined type, were observed with reobstruction or restenosis during follow-up. Among them, there were 24 lesions of the IVC (n = 12) and HV (n = 12). There was no significant difference between reobstruction of IVC and HV (12/58 vs 12/70; P = .516). The cumulative 1-, 3-, and 5-year primary patency rates of the HV were 96.7%, 90.0%, and 83.3%, respectively. The cumulative 1-, 3-, and 5-year primary patency of the IVC was 86.7%, 71.7%, and 68.3%, respectively. These patients were successfully managed by adequate anticoagulation and repeated IVC or HV endovascular intervention (Fig 3).

Univariate analysis of factors that may affect patency of the HV or IVC showed that the patients with splenomegaly (P = .097), substandard 12-month-long anticoagulation (P = .004), or etiology of membrane formation (P = .004)

Table. Clinical features of the 83 patients with primary Budd-Chiari syndrome (BCS)

Variable	Value
Age, years	40 ± 12.4
Male/female	55/28
Duration of symptoms, months	0.5-384
Comorbidities	
Hypertension	2 (2.4)
Diabetes	2 (2.4)
Behçet disease	1 (1.2)
Pulmonary embolism	2 (2.4)
Deep venous thrombosis	4 (4.8)
Smoking history	17 (20.5)
Drink history	15 (18.1)
Clinical presentation	
Encephalopathy	5 (6.0)
Stage 1	2 (2.4)
Stage 2	0 (0.0)
Stage 3	3 (3.6)
Stage 4	0 (2.4)
Ascites	37 (44.6)
Mild, <500 mL	20 (24.1)
Moderate, 500-3000 mL	8 (9.6)
Massive, >3000 mL	9 (10.8)
Abdominal pain or distention	51 (61.4)
Jaundice	49 (59.0)
Hepatomegaly	59 (71.1)
Splenomegaly, hypersplenism	56 (67.5)
Gastroesophageal bleeding varices	27 (32.5)
Thoracoabdominal wall distended veins	43 (51.8)
Lower extremity edema	45 (54.0)
Laboratory values	
Alanine transaminase, U/L	153 ± 91
Aspartate transaminase, U/L	53 ± 45
Total bilirubin, μmol/L	45.9 ± 29.0
Direct bilirubin, μmol/L	21.0 ± 15.6
Albumin, g/L	37.2 ± 5.8
Creatinine, μmol/L	75 ± 36
White blood cell count, 10^9/L	5.08 ± 2.87
Hemoglobin, g/L	123.9 ± 24.6
Platelet count, 10^9/L	121.8 ± 66.2
Prothrombin time, seconds	13.6 ± 1.8
International normalized ratio	1.22 ± 0.17
Child-Pugh score	
Class A	40 (48.2)
Class B	38 (45.8)
Class C	5 (6.0)
MELD score	8 ± 5
Clinical classification	
IVC	13/83 (15.7)
HV	25/83 (30.1)
IVC + HV	45/83 (54.2)
Pathologic type	
Thrombus	13/83 (15.7)
Membrane	22/83 (26.5)
Combined component	48/83 (57.8)

HV, Hepatic vein; *IVC,* inferior vena cava; *MELD,* Model for End-Stage Liver Disease.
Categorical variables are presented as number (%). Continuous variables are presented as mean ± standard deviation.

were more likely to relapse. Further multivariate analysis, especially for the IVC type, showed that etiology of membrane formation and substandard 12-month-long anticoagulation were independent determining factors for postoperative relapse or restenosis (Fig 4).

DISCUSSION

This study demonstrated the feasibility, strategy, and long-term results of a radical surgical recanalization through exposure of the entire IVC of the hepatic segment for BCS. Patients in this study had a history of failed endovascular treatment. Compared with TIPS, the radical surgical procedure, although more invasive, has an opportunity to re-establish hepatic outflow and partially restore liver function. As a decompression procedure, TIPS may palliate liver cirrhosis and prevent complications of portal vein hypertension with certain risks, including hepatic encephalopathy and restenosis. Because of high expense and donor scarcity, not all end-stage patients can afford liver transplantation. Radical surgery may be considered an alternative procedure to prevent disease progression.

A systematic review of 79 studies of BCS treatment demonstrated that according to the treatment modality, the median 1-, 5-, and 10-year survival rate was 93%, 83%, and 73% after intervention; 81%, 75%, and 72.5% after surgery other than liver transplantation; and 68.1%, 44.4%, and unavailable after medical therapy alone.[18] Compared with the review data, our survival rates of 1 year and 5 years were better than the reported average for surgery and as good as those of the intervention groups.

In practical terms, dozens of surgical recanalization methods for BCS are applied to restore blood flow of the HV or IVC and to improve the congested venous state. The common surgical methods include finger membranotomy through the right atrium, radical resection of membrane and thrombus, bypass operations, portosystemic shunting, and combined surgery

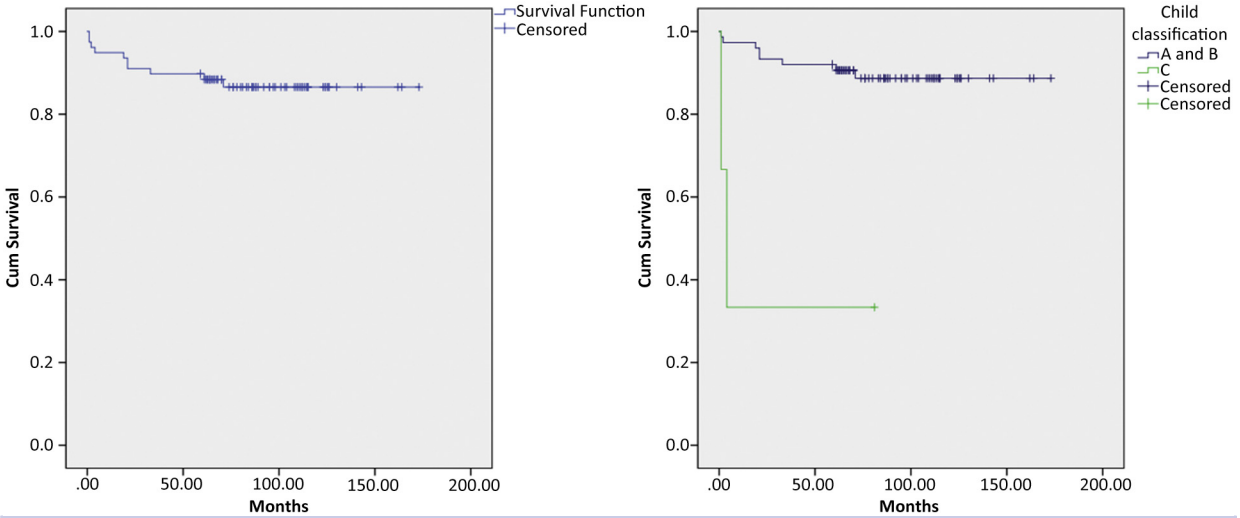

Fig 2. *Left*, Survival after surgical treatment. *Right*, Survival after surgical treatment between Child-Pugh A and B and Child-Pugh C scores.

(intervention plus bypass or shunt surgery).[1-3,15] However, the outcome of each surgical recanalization is slightly different as the indication is not identical.

Nonfulminant patients with thrombosis or compression and a high pressure in the infrahepatic IVC can be considered for surgical shunting. Orloff et al[19] described a group of patients with BCS who received a side-to-side portacaval shunt or mesocaval shunt therapy with no technical failure. The long-term survival of patients who received a side-to-side portacaval shunt was >95%, with complete resolution of ascites and no encephalopathy in 54 patients. All patients were followed up for 5 to 25 years, and histologic evaluation of the liver showed stable fibrosis in 52% of patients and no regressing cirrhosis. However, the common clinical problems are that congested hypertrophied caudate lobes often make the side-to-side portacaval shunt difficult to construct and that the long prosthetic grafts easily undergo long-term sclerotic changes to develop severe stenosis or thrombose despite routine anticoagulation.[19-22] Some reports also showed that there were no significant survival advantages for patients with severe liver dysfunction, causing an average 25% hospital or perioperative mortality.[22-24] European multicenter studies have demonstrated that the surgical shunt has been gradually replaced by TIPS because of a lower invasiveness and a relatively easier procedure for similar cases.[25,26]

Other procedures that emphasize direct removal of the lesion, such as finger membranotomy through the right atrium or radical resection of membrane and thrombus, showed some superiority in patency and survival. Xu and Dang[27] documented clinical effectiveness rates of 61.4%, 91.7%, and 90.4% in patients with BCS who were treated with finger membranotomy, endovascular intervention,

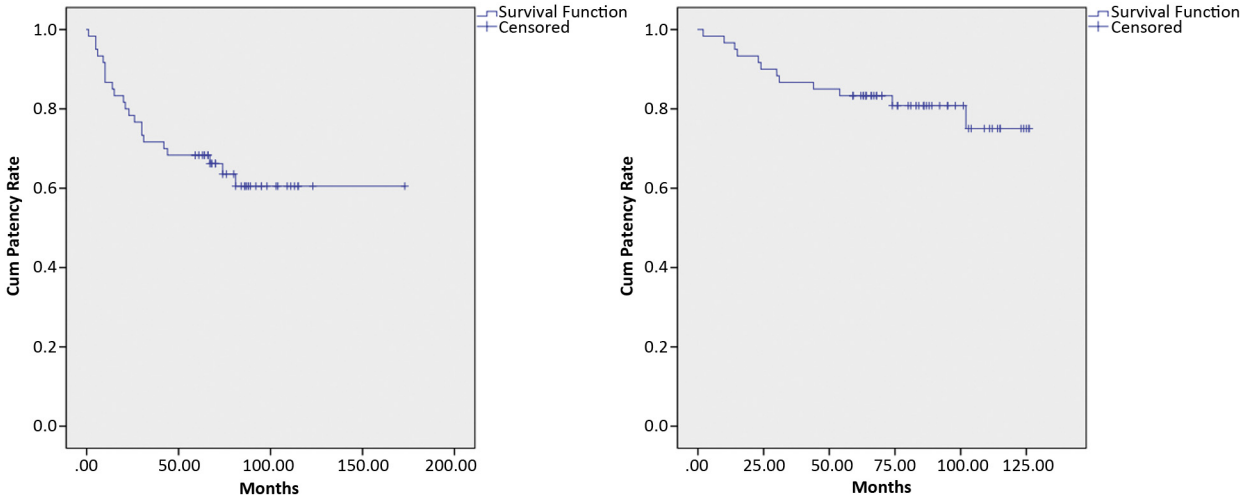

Fig 3. *Left*, Primary patency of the inferior vena cava (IVC) after surgical treatment. *Right*, Primary patency of the hepatic vein (HV) after surgical treatment.

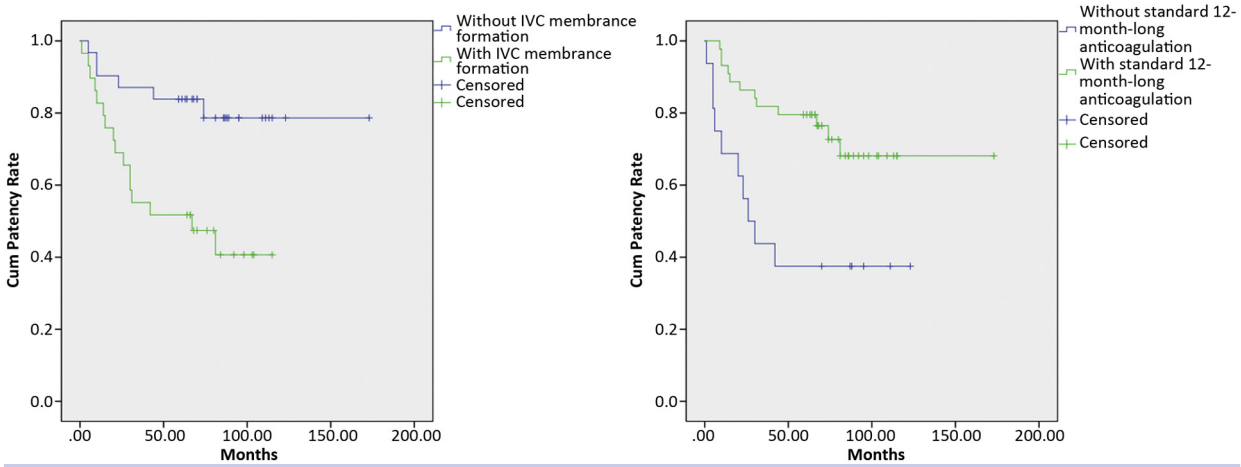

Fig 4. *Left*, Primary patency of the inferior vena cava (*IVC*) after surgical treatment with or without IVC membrane formation. *Right*, Primary patency of the IVC after surgical treatment with or without standard 12-month-long anticoagulation.

and membrane resection, and the recurrence rate was 38.4%, 8.3%, and 9.6%, respectively, which showed significantly better long-term effect in the resection group and interventional group than in the finger membranotomy group. They also suggested that patients with an extensive lesion, thick membrane, or recurrence after intervention should undergo membrane resection. A study of IVC thrombectomy or plasty with a pericardial patch performed in Japan reported that the 5- and 10-year primary patency rates of IVCs were 90.5% and 84.3%, whereas the 5- and 10-year survival rates were 89.8% and 70.7%, respectively.[28] Inafuku et al[28] also found that 32 patients (60.3%) in their study had histologic liver cirrhosis, and their cumulative survival rates were 93.3% at 5 years and 78.2 % at 10 years. They suggested that TIPS should be considered as a bridge to definitive surgery in case of fulminant or acute liver failure of BCS with liver cirrhosis.

Our study showed better outcomes than in other reports. We speculated that there may be four advantages. First, the improvement of our surgical method lies not only in the removal of either membrane or thrombosis more thoroughly but also in the direct view through exposure of the entire IVC of the hepatic segment and HV outflow tract to ensure the resection of both IVC and HV lesions at the same time. Second, compared with the "skip" of the hepatic sinus function of TIPS, the reconstructed blood flow of portal vein-hepatic sinus-HV of our operation is more physiologic for humans. In addition, the cardiopulmonary bypass machine is able to pump the blood into the body circulation to maintain blood volume when the IVC is incised. Last, a direct operative view through exposure of the entire IVC of the hepatic segment and HV outflow tract is helpful to observe the location of the lesion at the IVC and HV. Furthermore, we can also easily collect relatively intact samples and remove a fractured IVC stent once it is implanted or local tumor involving the ICV through our operation.

The most common major complication of our radical surgical procedure was massive hemorrhage. The most frequent reason for difficulty in obtaining hemostasis intraoperatively was a coagulation disorder secondary to hepatic dysfunction as well as thrombocytopenia secondary to hypersplenism; thus, detailed estimation of blood coagulation function is of much importance. Besides, severe liver laceration was relatively less common. In our center, all surgeons had experience in surgical recanalization methods, such as finger membranotomy through the right atrium, radical resection of membrane and thrombus, and bypass surgery.

Another important question is how risk factors affect the outcome of surgical therapy for BCS. Multiple studies have demonstrated that encephalopathy, bilirubin concentration, creatinine level, and ascites might be more closely associated with the survival of patients undergoing surgical therapy for BCS, which helps stratify the prognosis of BCS and identifies candidates for more invasive treatment modalities in a timely fashion.[15] Li et al[29] retrospectively analyzed 219 patients with postoperative recurrence and also found that age, total bilirubin concentration, and severity of liver function were the independent recurrent indicators contributing to thrombosis or subsequent fibrous obstruction. In addition, an underlying prothrombotic disorder or an established risk factor for venous thrombosis is found in most patients, which may contribute to restenosis. However, thrombotic risk factors are different between China and Western countries, such as the low incidence of myeloproliferative neoplasms with a JAK2 V617F mutation (3%) or V Leiden mutation (0%) and the similar incidence of hyperhomocysteinemia (40%) or anticardiolipin antibodies (16%).[1,14,30,31]

We analyzed the survival and recurrence rates in our study. Univariate analysis showed some new findings as well as some general conclusions that were drawn by other reports. Age, encephalopathy, and ascites were also the important factors affecting surgical mortality, and the uncommon risks, such as diabetes and splenomegaly, were new significant related factors. However, after multivariate analysis of these factors, Child-Pugh C score was determined to be the only independent risk factor for surgical survival. The cumulative 1-, 3-, and 5-year survival rates of patients with class A and B Child-Pugh scores were 97.3%, 92.0%, and 90.6%, respectively; however, those of Child-Pugh class C scores were 66.7%, 33.3%, and 33.3%, respectively.

The analysis of restenosis or recurrence in the IVC or HV showed that the patients with oral anticoagulation of <12 months or etiology of membrane formation were more likely to relapse. Specifically, the cumulative 1-, 3-, and 5-year patency rates of patients without membrane formation were 90.3%, 87.1%, and 83.9% compared with 82.8%, 55.2%, and 51.7% for those with membrane formation. The cumulative 1-, 3-, and 5-year patency rates of patients with standard oral warfarin were better than those of patients without (93.2%, 81.8%, and 79.5% vs 68.8%, 43.8%, and 37.5%). We suggest that all postoperative patients receive long-term (perhaps lifelong) standard anticoagulation unless it is contraindicated, starting with intravenous heparin, then warfarin, maintaining the international normalized ratio around 2.5. For the other factor of membrane formation risk, we speculate that the postoperative residual membrane increases the risk of subsequent thrombosis, especially under the basic condition of hypercoagulability. In addition, hepatomegaly may contribute to false restenosis of the IVC or HV because of compression.

CONCLUSIONS

Compared with shunting procedures, radical surgery for BCS has the advantage of physiologic restoration of hepatic outflow and use of liver function to a maximum extent. Our radical procedure can provide a direct-view operative field through exposure of the entire hepatic IVC and hence better results on rates of survival and recurrence. For BCS patients with combined IVC and HV involvement or repeated failure after endovascular therapy, we recommend the direct-view radical operation as a reasonable choice. The limitation of this study is that it is a single-center retrospective research without randomization of different treatment methods. Further studies with larger scale and long-term follow-up are definitely necessary to accumulate more convincing evidence.

AUTHOR CONTRIBUTIONS

Conception and design: XiZ
Analysis and interpretation: QL, TZ
Data collection: DW, WL, XuZ
Writing the article: QL, TZ
Critical revision of the article: DW, WL, XuZ, XiZ
Final approval of the article: QL, TZ, DW, WL, XuZ, XiZ
Statistical analysis: TZ
Obtained funding: XiZ
Overall responsibility: XiZ
QL and TZ contributed equally to this article and share co-first authorship.

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