

Surgical treatment of 1360 cases of Budd-Chiari syndrome: 20-year experience

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BACKGROUND: Budd-Chiari syndrome (BCS) is a disease caused by blood flow obstruction of the main hepatic veins (MHVs) and/or the outlet of the inferior vena cava (IVC), characterized by retrohepatic portal hypertension (PHT) and/or IVC hypertension. In the past decade, over 3000 cases of BCS have been reported in China. This study was to sum up our 20-year experience in surgical treatment of BCS and to investigate its pathological classification and principles of surgery.

METHODS: The data from 1360 BCS patients were analyzed retrospectively.

RESULTS: Four types (6 subtypes) were classified according to IVC angiography and hepatovenography: type Ia (594 patients), type Ib (123), type II (292), type IIIa (237), type IIIb (112), and type IV (2). Surgical procedures included: improved splenopneumopexy (265 cases), finger or balloon membranotomy (407), radical resection of membrane and thrombus (275), IVC bypass (88: cavocaval transflow 71 cases, and cavoatrial transflow 17 cases), mesocaval C-shape shunt (192), splenocaval shunt (32), splenoatrial shunt (23), splenojugular shunt (57), mesoatrial shunt (8), and combined methods (6), including plenal-cavoatrial shunt (4), and mesocavoatrial shunt (2), spleno-renal shunt (4), mesojugular shunt (2), and other methods (1). The perioperative death rate and the complication rate after operation was 3.09% (42/1360) and 14.8% (201/1360) respectively. 885 cases were followed up from 9 months to 15 years (average 6.8 ± 1.2 years). The 791 (89.4%) of 885 patients were successfully treated, 61 patients (6.89%) had a recurrence, and 33 died.

CONCLUSION: Surgical treatment of BCS is dependent on a correct diagnosis and classification of the disease.

(*Hepatobiliary Pancreat Dis Int* 2004; 3: 391-394)

KEY WORDS: Budd-Chiari syndrome; diagnosis; surgical treatment

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Introduction

Budd-Chiari syndrome (BCS) represents a syndrome with retrohepatic portal hypertension and/or inferior vena cava hypertension which results from the obstruction of the main hepatic veins (MHVs) and/or the retrohepatic inferior vena cava (RHIVC).^[1-3]

BCS was reported scarcely before the 1980s and the cases of this syndrome reportedly increased due to the development of medical technology, especially the development of imaging medicine. This disease is a severe benign disease, with a poor prognosis. Medication of this disease can not get good result but dozens of surgical therapies can produce good result according to the different pathologic types of the disease. Since we treated the first BCS patient at this hospital in May 1983, a total of 1360 patients with BCS have been treated surgically with success.

Methods

Patients

In the 1360 patients, 833 were male, and 527 female, aged from 4.5 to 71 years (average 33.2 ± 3.6 years). The history of BCS in these patients ranged from 1 month to 18 years. All the patients showed symptoms and signs of portal hypertension (PHT) and/or hypertension of the inferior vena cava (IVC).^[1-3] 1292 patients demonstrated inertia, 1284 abdominal distension, 662 decreased appetite, 122 abdominal pain, 116 jaundice, 1124 hepatomegaly, 821 chest or abdominal wall varix, 914 ascites, 868 leg edema or pigmentation, 162 upper gastrointestinal hemorrhage (hematemesis or melena), and 683 splenomegaly. Liver function was classified by Child-Pugh grading: grade A in 831 patients, B in 408, C in 121. Histological and pathological examination showed expanded liver antrum in 136 patients with atrophic liver cells and necrotic or fibrosed cells around the center of lobules of the liver. All patients diagnosed by IVC angiography (1108 patients), percutaneous hepatovenography or color Doppler (252) were classified according to reported references:^[1,4,5] type Ia (594 patients); type Ib (123); type II (292); type IIIa

(237); type IIIb (112); and type IV (2).

Statistical analysis

We used the SPSS 10.0 statistical software package to deal with the data and the chi-square test was used to compare the quantitative data.

Results

The criteria of effects of the treatment were defined.^[6] Good effect: no clinical symptoms, ascites, chest and abdominal wall varix, and leg edema were found, nor

the liver could be touched under the ribbon arch. Improved effect: chest and abdominal wall varix almost disappeared, the liver shrunk, and ascites reduced obviously. No effect: the clinical symptoms remained or worsened. The therapeutic methods and their results in this group are shown in Table 1. The perioperative mortality was 3.09% (42/1360) and the incidence of complication was 14.8% (201/1360) (Table 2).

885 patients were followed up for 9 months to 15 years, averaging 6.8 ± 1.2 years. The means of follow-up included telephone, letter and out-patient visit. The effective rate of the treatment was 89.4% (791/885), but there were 33 deaths, due to hepatic encephalopathy 1 to 3 years after operation (6), upper gastrointestinal hemorrhage 1 to 5 years after operation (7), hepatic carcinoma after operation (4), renal failure after operation (3), and others cause other than BCS operation.

Table 1. The effect and clinical therapy methods

Therapy method	Cases	Effect			
		Good	Improved	No effect	Death
Improved splenopneumopexy	265	68	179	12	6
Kimura's rupture or Foley's tube dilation	407	296	79	27	5
Radical membrane resection and thrombus extracting	275	166	76	20	13
Cavocaval transflow	71	60	7	2	2
Cavoatrial transflow	17	10	3	2	2
Mesocaval C shunt	192	116	56	18	2
Splenicaval shunt	32	22	8	2	0
Splenoatrial shunt	23	7	6	6	4
Splenojugular shunt	57	39	10	7	1
Mesoatrial shunt	8	1	1	2	4
Spleno renal shunt	4	1	2	1	0
Spleno cavoatrial shunt	4	0	2	1	1
Mesocavoatrial shunt	2	0	1	0	1
Mesojugular shunt	2	0	0	2	0
Other method	1	0	0	0	1
Total	1360	786	430	102	42
Percentage (%)		57.79	31.62	7.50	3.09

Table 2. 201 BCS patients with complications and results

Complication	Cases	Results	
		Cured	Death
Hydrothorax	68	68	0
Acute heart failure	29	23	6
Hemorrhagic shock	27	17	10
Acute liver failure	13	5	8
Upper gastrointestinal hemorrhage	12	9	3
Acute respiratory function failure	9	7	2
Pulmonary embolism	5	1	4
Chyle leakage	21	21	0
DIC	5	1	4
MODS	5	0	5
Pericarditis	4	4	0
Infection of artificial vessel	3	3	0
Total	201	159	42
Percentage (%)	201/1360 (14.8)	159/1360 (11.7)	42/1360 (3.1)

Discussion

In western countries, BCS are mainly Chiari diseases characterized by hepatic veins obstruction after thrombosis. Since the membranous obstruction of the inferior vena cava (MOIVC) in Asian countries and South Africa is similar to Chiari disease, BCS is considered extensively a syndrome with retrohepatic PH and/or IVC hypertension caused by blood stream obstruction in MHVs outlets and/or RHIVC. China, Japan, India, South Africa are countries with a high incidence of BCS.^[1,3] Reports of the past decade have shown that over 3000 cases were treated in China's Henan, Shandong, Jiangsu and Anhui provinces.^[1,5,7] Researchers found that pregnancy, dysfunction of blood coagulation, infection, oral contraceptives, trauma, and tumor may cause BCS.^[2,8]

Pathological classification

With complicated condition and different types, BCS is not classified uniformly and pathologically. In 1970, Hirooka^[9] classified BCS into 7 types including 10 subtypes, but it was inconvenient to use clinically. Pei-Qin Xu and coworkers^[5,10] classified BCS into 4 types (6 subtypes): type Ia for membranous obstruction of the vena cava (MOVC) without thrombus under membrane and MHVs are patent or partly patent; type Ib for MOVC with thrombus under membrane, and MHVs are patent or partly patent; type II for segmentally narrowed IVC and segmentally obstructed MHVs; type IIIa for segmentally obstructed IVC (<2 cm) and obstructed MHVs, with right inferior posthepatic vein compensatively swollen; type IIIb for segmentally obstructed (≥ 2 cm) and obstructed MHVs without compensatively swollen veins in the third portion of the portal hepatis; type IV for whatever the type narrowing or obstruction of the superior vena cava. This method of classification is reasonable and practical, proved by our clinical practice. Moreover, using our own diagnostic

standards for severe BCS all patients with acute or chronic progress proved clinically or radiologically were considered as having severe BCS if one of the following conditions was met:^[5,11] (1) refractory ascites and abdominal pressure ≥ 2.7 kPa (20 mmHg); (2) oliguria or anuria; (3) marked liver dysfunction, prothrombin time longer than 50%, inverted rate of albumin/globulin and serum bilirubin >34.2 mmol/L; (4) associated hepatic encephalopathy; and (5) upper gastrointestinal hemorrhage.

Treatment methods

A dozen of surgical methods for BCS,^[12] including operations for membrane, averting flow methods, shunts and other combined operations. The main purposes of these operations^[13] are to release obstruction, to recover blood stream in the MHV and IVC, to decompress with shunts or averting flow methods, to relieve congestion of the liver and IVC system, to control bleeding, to remit hypersplenism, to eliminate ascites, and to protect the liver. Some scientists^[14,15] reported the use of transjugular intrahepatic portosystemic shunt (TIPS) in the treatment of BCS, but we consider that the principles of TIPS are contradictory to the therapeutic principles of BCS. The basic pathologic change of BCS is the occlusion of the outlet of the hepatic vein, i. e. the outlet of the hepatic vein and retrohepatic IVC are blocked. How can we use TIPS to treat BCS? If we choose to puncture the outlet of the hepatic vein and redo the outlet, why it is called "intrahepatic portosystemic shunt?" Strictly the so-called TIPS for the treatment of BCS should be named as transjugular hepatic venoplasty/veno-reconstruction shunt (THVS). Therefore, TIPS for the treatment of BCS still disputable.^[5] As to liver transplantation, it is only suitable for end-stage liver disease associated with BCS^[2] in consideration of economic burden of the patient.

Operations for lesioned membrane

Despite Kimura's finger rupture,^[16] Foley's tube dilation,^[17] radical membrane resection and thrombus extracting,^[18] we used radical membrane resection combined with thrombus extracting under direct vision and at normal temperature. In treating 480 patients with membranous BCS we found the effective rates of the rupture group, interventional group and radical group were 61.4%, 91.7% and 90.4% respectively and their recurrence rates were 38.6%, 8.3% and 9.6%. The long-term effects of the interventional group and the radical group were better than those of the rupture group ($P < 0.05$), and the recurrence rates were lower than that in the rupture group ($P < 0.05$).^[6] In a word, BCS patients suitable for Kimura's finger rupture should take Foley's tube dilation as the first choice but for those with thrombus beneath the membrane, large lesion, thick and tough membrane, leaning membrane, and re-

currence after interventional treatment, radical membrane resection combined with thrombus extracting is advisable.

Transflow or IVC bypass

Artificial vessel is used to recover the blood flow between the lower and upper parts of the IVC or right atrium close to the lesion including the cavocaval transflow and cavoatrial transflow.^[19,20] Comparison of the two methods showed that cavocaval transflow is advantageous in lowering the length of artificial blood vessel and the rate of thrombosis, changing intrapericardium transflow into extra pericardium one, decreasing the incidence of pericardial tamponade and pericarditis, simplifying operative procedures and lessening injury. In this group, 71 patients showed cavocaval transflow and 17 cavoatrial transflow, with a complication rate of 9.86% (7/71) and 41.18% (7/17) respectively ($P < 0.05$, $\chi^2 = 9.07$).

Shunts^[21-23]

Shunts transfer the blood flow of the portal vein to the systemic circulation partly or totally, reduce the symptoms of PHT decompress PHT, and prevent upper gastrointestinal bleeding. They include mesocaval C shunt, splenojugular shunt, splenocaval shunt,^[13] mesoatrial shunt, splenorenal shunt, and others. Splenojugular shunt could be used as the first choice for severe BCS.^[11] In 43 patients undergoing shunts, 19 had mesoatrial shunt reported by Slakey et al,^[8] and 24 mesocaval C shunt with a perioperative mortality of 31.58% (6/19) and 16.67% (4/24) respectively. The 33 patients were followed up, and one patient who had had mesoatrial shunt had recurrence of obstruction. In another group of 60 BCS patients, 32 had portacaval shunt with a perioperative mortality of 3% but 94% of them lived for 3.5-27 years after operation (without ascites and normal liver function in 31 patients), and 5 of 8 patients who had had mesoatrial shunt died of upper gastrointestinal bleeding owing to the blocking of artificial vessels.^[10] In our 8 patients who had had mesoatrial shunt, 2 demonstrated good results, and 4 died. Thus we advocate to give up mesoatrial shunt. Based on Akita's method, we reduced two incisions in the chest to one in the abdomen, and added an epiploon to enclose the lung.^[24] With this method we treated 202 PHT patients, with an effective rate of 93.1% after follow-up for 7 months to 12 years. This method is applicable to the patients without intractable ascites and lung and heart dysfunction.

Combined operation

Combined operation such as side-to-side portacaval shunt and cavoatrial shunt is designed to transfer two thirds blood volume of the lower part circulation and the total portal blood volume. Ten BCS patients were reported to undergo this operation, with the decreased

pressure between the IVC and right atrium from 195 to 22 mmH₂O. Four to 16-year follow-up showed all patients were alive after patent shunt.^[2] Mesocaval and cavoatrial shunts for 14 BCS patients in 4 years showed a short-term patent rate of blood vessels for 86% and that of mesoatrial shunt for 93%. It needs further observation since the follow-up time is short.^[25]

Principles for choosing operative procedures

Different methods should be used according to different types of lesions and hemodynamic changes of patients so as to decrease the perioperative death rate and postoperative complications.^[2,4,5,26] In type Ia patients, the interventional Foley's tube dilation should be the first choice. In some patients with a large lesion, thick and tough membrane, leaning membrane and postoperative recurrence, radical membrane resection is advisable. In type Ib, radical membrane resection and thrombus extracting or IVC bypass should be the first choice. In type II, different shunts (mesocaval shunt or splenocaval shunt) are chosen according to hemodynamic changes. In patients with splenomegaly, partial splenectomy or improved splenopneumopexy can be used as the first choice. In type IIIa, Foley's tube dilation and stent placement, radical membrane resection, and thrombus extracting or IVC bypass are applicable. In type IIIb, splenojugular shunt, splenoatrial shunt or combined operation are feasible.

Competing interest

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

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Received March 3, 2004

Accepted after revision June 31, 2004