

# Diagnosis and management of severe Budd-Chiari syndrome

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**OBJECTIVE:** To assess the diagnostic standards and treatment of severe Budd-Chiari syndrome (BCS).

**METHODS:** The clinical data of 126 patients with severe BCS treated from November 1994 to June 2001 at our hospital were retrospectively analyzed. Percutaneous transhepatic recanalization and dilation and/or stent placement of the main hepatic vein was performed in 10 patients. Mesocaval C type shunt with artificial graft was performed in 68 patients, splenojugular shunt in 33, mesojugular shunt in 1, and mesocaval shunt or improved splenopneumopexy after percutaneous intraluminal angioplasty and stent placement of the inferior vena cava in 14.

**RESULT:** Six patients died during perioperation. In 120 patients followed up for 6 months to 7 years, 89 had excellent results and 31 good results.

**CONCLUSIONS:** Diagnostic standards of severe BCS are suggested. Proper treatment should be used according to the pathological changes of the inferior vena cava and main hepatic veins.

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**Key words:** Budd-Chiari syndrome, severe; diagnosis; therapy

## Introduction

Budd-Chiari syndrome (BCS) characterised by outflow obstruction of the hepatic veins and terminal portion of the inferior vena cava (IVC) is considered as a kind of portal hypertension (PHT) often associated with IVC hypertension.<sup>[1]</sup> In patients of western countries, the lesion is often located in hepatic veins (HV), but in China, Japan, India and South Africa, membranous occlusion in the IVC is easier to see.<sup>[2,3]</sup> Owing to the improvement of diagnosis and management, a lot of cases were diagnosed.<sup>[4]</sup> In India, the rate of portal hypertension was 7%–9%.<sup>[5]</sup> Physical therapy would produce a poor prognosis,<sup>[6–8]</sup> and the natural death rate could reach 22%–42% one year after diagno-

sis.<sup>[9]</sup> Because complicated conditions of BCS, especially severe BCS, improper therapy will lead to poor prognosis. From November 1994 through June 2001, 126 patients with severe BCS were treated successfully at our hospital with operation or interventional therapy or both in combination.

## Methods

Of the 126 patients, 71 were male and 55 female, aged from 18 to 45 years (mean 29.6). Their clinical manifestations are listed in Table.

Oliguria (urine within 400 ml per day) and anuria (urine within 100 ml per day) were noted in some patients. The patients with conjunctiva with jaundice were characterized by a level of serum bilirubin higher than 34.2 mmol/L, and some patients manifested with anemia, tiredness, and poor appetite. Liver enlargement was felt in different degrees, even at the umbilicus line. The spleen was enlarged in a mild to moderate degree, while moderate to large amount of ascites, and abdominal pressure from 20 mmHg to 38 mmHg were

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**Table.** Clinical manifestations of 126 patients with severe BCS

Manifestations	n	%
Liver enlargement	126	100.00
Ascites	126	100.00
Leg edema	71	56.35
Spleen enlargement and hyperfunction	68	53.97
Oliguria or anuria	66	52.38
Chest and abdominal superficial varices	48	38.10
Conjunctiva jaundice	22	17.46
Upper gastrointestinal bleeding (hematemesis or melena)	21	16.67
Varices or pigmentation of leg	19	15.08
PT prolonging over 50%	18	14.29
Overtured albumin/globulin	13	10.32
Umbilical hernia	6	4.76
Female infertility	6	4.76
Hepatic encephalopathy	2	1.59

PT: prothrombin time.

detected.

### Diagnosis and typing

All patients selected after B-type ultrasound or color Doppler were confirmed by IVC angiography and/or percutaneous transhepatic venous angiography. According to Pei-Qin Xu's typing,<sup>[10]</sup> 81 patients belonged to type II and 45 patients type III.

### Treatment

#### Preoperative treatment

Preoperative treatment aimed at protection of hepatic function, improvement of general condition by supportive treatment, and correction of hydro-electrolyte and base-acid disturbance was applied to all patients. In patients with a large amount of ascites complicated by abdominal compartment syndrome (ACS),<sup>[11]</sup> condensed ascites transfusion and other methods were used to decrease abdominal pressure, and to relieve the symptoms of ACS. In this group, 48 patients received condensed ascites transfusion one to three times, the largest amount being 10 000 ml in one patient.

#### Treatment method

In 10 patients treated interventionaly in this group, percutaneous transhepatic recanalization

and dilation and/or stent placement was performed. 102 patients received operation including mesocaval C type shunt (68 patients), splenojugular shunt using the right internal jugular vein, otherwise the left (33), and mesojuglar shunt (1). In 14 patients who received operation and interventional therapy, Seldinger's technique was used to dilate the IVC and place stent for the purpose of mesocaval C type shunt or improved splenopneumopexy.

### Results

In this group, 6 patients died postoperatively: three patients receiving mesocaval C type shunt died of liver failure (2) and DIC (1); 2 patients receiving splenojugular shunt died of liver failure; and one patient having combined operation and interventional therapy died of liver failure. The remaining patients were alive. 120 patients were followed up for 6 months to 7 years, and no patient had recurrence of the syndrome or worsening. Eighty-nine patients (74.2%) showed disappearance of clinical symptoms and ascites as well as normalization of liver size. 31 patients (25.8%) showed disappearance of clinical symptoms, reduced size of the liver, and reduction of ascites. Two patients were complicated by interrupted hepatic encephalopathy which was relieved by control of protein diet and improvement of liver function. In 86 patients who were checked by color Doppler after operation, 68 patients showed potency of vascular graft. Two patients receiving splenojugular shunt suffered thrombosis caused by graft infection one year after operation, and subsequent operative removal of grafts showed no recurrence of symptoms.

### Discussion

#### Diagnostic criteria for severe BCS

Our experiences have shown that BCS patients who are confirmed clinically or radiographically should be considered as having severe BCS if they meet the following conditions: (1) stubborn ascites and abdominal pressure no less than 20 mmHg; (2) oliguria or anuria; (3) serious liver dysfunction,

prothrombin time (PT) longer than 50%, overturned albumin/globulin level, serum bilirubin level over 34.2 mmol/L; (4) being or once complicated by hepatic encephalopathy; (5) being or once complicated by upper gastrointestinal bleeding.

### Preoperative management of severe BCS

Severe BCS patients often have enlarged liver, jaundice, a plenty of ascites, hypolipoproteinaemia, dyspnea, and oliguria or anuria. The patients with ACS who are intolerable to operation are likely to increase the incidence of postoperative complications and affect the prognosis if they are not treated properly before operation. In this group, the patients having a plenty of ascites especially those with ACS were subjected to transfusion of condensed ascites. As a result their abdominal swelling and dyspnea were relieved, and urine volume and serum albumin level increased to allow an operation. In addition, improving the general condition of patients by protecting liver function and prescribing supportive treatment, and correcting the disturbance of hydroelectrolyte and acid-base balance can improve patients' general condition and reduce the incidence of postoperative complications. Six patients with liver function of Child B to Child C before operation died from liver failure and other complications.

### Choice of operation

The main purpose of operation for BCS is to remove the obstruction of the hepatic veins and IVC or construct bypass to lower the pressure of the hepatic veins and IVC.<sup>[12]</sup> For patients with severe BCS, operation should be selected according to the type of the disease, but fundamental therapy is to relieve portal hypertension.

In patients with a narrow segment ( $\geq 2$  mm) of the IVC, dilation of the main hepatic vein and membrane obstruction in the outlet, percutaneous transhepatic recanalization, and dilation or stent placement<sup>[13]</sup> could be performed to lower the pressure of the IVC and prevent gastroesophageal varices.

In those patients with a narrow segment ( $\geq 5$

mm) of the IVC, inner pressure  $< 19$  mmHg, complete obstruction of hepatic veins, and no dilation of short hepatic veins in the third porta hepatis, mesocaval C shunt<sup>[14]</sup> may be performed, and if the IVC diameter  $< 5$  mm, inner pressure  $\geq 19$  mmHg, splenojugular shunt or spleen-atrium shunt is advisable.<sup>[15]</sup> We advocate the combined treatment to deal with the above two conditions by using percutaneous IVC recanalization and dilation and stent placement, followed by mesocaval shunt or improved splenopneumopexy even in patients with mild ascites.<sup>[10]</sup>

If both the portal vein system and IVC system have a high pressure and patients have distinct symptoms, combined shunts or IVC bypass outside the pericardium is feasible.<sup>[16]</sup> Liver transplantation is applicable to patients with end-stage liver disease.<sup>[17]</sup>

### 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.

### References

- 1 Okuda K. Review: membranous obstruction of the inferior vena cava (obliterative hepatocavopathy). *J Gastroenterol Hepatol* 2001;16:1179-1183.
- 2 Dilawari JB, Bamberey P, Chawla Y, et al. Hepatic outflow obstruction ( Budd-Chiari syndrome): experience with 177 patients and a review of the literature. *Medicine* 1994;73:21-36.
- 3 Kohli V, Pande GK, Dev V, et al. Management of hepatic venous outflow obstruction. *Lancet* 1993;342:718-722.
- 4 Xu PQ. A 20-year experience with surgical treatment of Budd-Chiari syndrome (abstract). 4th international congress on the Budd-Chiari syndrome. Beijing China, October 2002.
- 5 Behera A, Menakuru S, Singh K, et al. Treatment of Budd-Chiari Syndrome with inferior vena caval occlusion by mesoatrial shunt. *Eur J Surg* 2002;168:355-359.
- 6 McCarthy PM, Van Heerden JA, Adson MA, et al. The Budd-Chiari syndrome: medical and surgical management of 30 patients. *Arch Surg* 1985;120:657-662.

- 7 Slakey DP, Klein AS, Venbrux AC, et al. Budd-Chiari syndrome; current management options. *Ann Surg* 2001;233:522-527.
- 8 Greenwood LH, Irizarry IM, Hallet JW, et al. Urokinase treatment of Budd-Chiari syndrome. *AJR* 1983; 145:1057-1059.
- 9 Hadengue A, Poliquin M, Vilgrain V, et al. The changing scene of hepatic vein thrombosis; recognition of asymptomatic cases. *Gastroenterology* 1994; 106: 1042-1047.
- 10 Xu PQ, Zhao YF, Zhang SJ. Surgical therapy of Budd-Chiari syndrome; report of 528 cases. *J Henan Med Univ* 1998;33:123-125.
- 11 Lu XB, Xu YJ, Ma XX, et al. Management of Budd-Chiari syndrome complicated with abdominal compartment syndrome. *Chin J Gen Surg* 2001;10:166-168.
- 12 Okuda K. Obliterative hepatocavopathy-inferior vena cava thrombosis at its hepatic portion. *HBPD Int* 2002;1:499-509.
- 13 Li TX, Hang XW, Ma WZ, et al. The investigation of interventional therapy for different of Budd-Chiari syndrome. *Chin J Radiol* 1999;33:181-184.
- 14 Ma XX, Zhao YF, Dang XW, et al. Mesocaval C-shunt with artificial vascular graft plus portaazygous disconnection in the treatment of Budd-Chiari syndrome. *J Zhengzhou Univ* 2002;37:564-566.
- 15 Xu P, Zhao L, Ma X. Management of severe Budd-Chiari syndrome by spleen-internal jugular venous shunt. *Zhonghua Wai Ke Za Zhi* 1998;36:290-291.
- 16 Xu P, Zhao L, Zhang SJ. Results of inferior cavocaval shunt in Budd-Chiari syndrome. *Chin J Gen Surg* 2000;15:210-212.
- 17 Orloff MJ, Daily PO, Orloff SL, et al. A 27-year experience with surgical treatment of Budd-Chiari syndrome. *Ann Surg* 2000;232:340-352.

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