

# Treatment of membranous Budd-Chiari syndrome; analysis of 480 cases

Pei-Qin Xu and Xiao-Wei Dang

Zhengzhou, China

**BACKGROUND:** Budd-Chiari syndrome (BCS) presents a kind of disease resulted from the occlusion of hepatic vein and/or the intrahepatic inferior vena cava. Its different pathological types were proposed. According to our experience, the membranous type takes a large part of it, and we tried to explore the best treatment of membranous BCS through the analysis of 480 cases retrospectively.

**METHOD:** The operative results of 480 patients with membranous BCS were analysed retrospectively.

**RESULTS:** Patients after Kimura's finger rupture, interventional treatment and membrane resection were followed up with rates of 84.62%, 86.55%, and 87.37%, respectively. The effective rates of the three methods were 61.4%, 91.7%, and 90.4%, respectively, and the recurrence rates of the disease after the 3 procedures were 38.6%, 8.3% and 9.6%, respectively. The long-term effects of interventional treatment and resection were significantly better than those of Kimura's finger rupture ( $P < 0.05$ ).

**CONCLUSION:** Balloon dilatation is the choice for membranous BCS. Patients with extensive lesion, thick membrane or recurrence after percutaneous transhepatic angiography should undergo membrane resection.

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**KEY WORDS:** hepatic vein thrombosis; balloon dilatation; vascular surgical procedures; Budd-Chiari syndrome

## Introduction

Budd-Chiari syndrome (BCS) is divided into different types according to the type, location and form of its obstruction.<sup>[1]</sup> In China, membranous obstruction is very common as that was reported in 528 BCS patients (1983 to 1997), 284 patients (53.79%) had membranous obstruction of the inferior vena cava

(MOVC).<sup>[2]</sup> By the end of 1990, we treated membranous BCS with Kimura's finger rupture through the atrium dextrum.<sup>[3]</sup> After 1990 we used interventional balloon dilatation. In 1993 we created a new method to treat membranous BCS with which we successfully resected membrane at normal temperature and direct vision. We analyzed retrospectively the long-term effects and recurrence rates of 480 membranous BCS patients who had been treated at our hospital from May 1983 to June 2000 in an attempt to explore the best therapy of membranous BCS.

## Methods

### Patients

Four hundred and eighty patients with membranous BCS had symptoms and signs of portal hypertension (PHT) and hypertension of the inferior vena cava (IVC). According to the report,<sup>[1]</sup> all of the patients were diagnosed by ultrasound or color Doppler primarily and proved by IVC and percutaneous transhepatic venous angiography. The 480 patients were divided into three groups: 52 patients who were treated with finger rupture via an atrium dextrum (rupture group) (33 men and 19 women, aged from 14 to 51 years, mean 28.5); 238 patients who were treated with interventional balloon dilatation (intervention group) (137 men and 101 women, aged from 18 to 59 years, mean 31.7); 190 patients who were treated with radical membrane resection (resection group) (110 men and 80 women, aged from 15 to 46 years, mean 29.1). The patients in the rupture group were treated before 1990, and those in the interventional group and resection group were selected according to the criteria reported. There were no statistical differences among sex, age and other aspects in the three groups.

### Treatment methods

#### Rupture group

Under general anesthesia, the patients laying on the left side underwent an intercostal incision of the right fourth spatium into the thoracic cavity. The pericardium was cut out, followed by a pursestring suture in the auricula dextra, which was scissored in side of the pursestring subsequently. The right index finger of the opera-

**Author Affiliations:** Department of General Surgery, First Affiliated Hospital of Zhengzhou University, Zhengzhou 450052, China (Xu PQ and Dang XW)

**Corresponding Author:** Pei-Qin Xu, MD, Department of General Surgery, First Affiliated Hospital of Zhengzhou University, Zhengzhou 450052, China (Tel: 86-371-6964308; Email: vl\_institute1086@163.com)

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tor was inserted into the IVC superior to the diaphragm through the nick to search and rupture the membrane. When it was satisfied, the finger was moved out and the suture was tightened. A pipe to draw liquid was placed in the thoracic cavity and the nick was closed.

#### **Interventional group**

Seldinger's technique was used to perform IVC angiography and measure the pressure. A balloon was inserted through the membrane and dilated for angiography. If necessary a stent was placed and angiography was performed again. If there was membranous obstruction of the main hepatic veins, percutaneous transhepatic venous angiography was performed. When the leading filament went through the membrane, the balloon was dilated for angiography.

#### **Resection group**

Under general anesthesia, the patient laying on the left side received an intercoastal incision of the seventh spatium into the thoracic cavity and the pericardium was sheared upright just in the right backward of the phrenic nerve. A belt was placed to obstruct the IVC through the pericardium (near-heart side) and uncover the retro-hepatic IVC completely. The belt was tightened to cut the right backward of the IVC at the membranous location upright. The index finger of the operator was inserted into the distal heart side of the IVC via dilation of the membrane in the outlet of the hepatic veins. If there was secondary thrombosis under the membrane in the IVC, the finger was taken out, while inserting a Foley tube into the distal side of the IVC after prompt incision. Twenty ml heparin saline was injected into the balloon, and then the tube was moved toward the head side to suck out the thrombus together with blood stream. At the same time, enlarged balloon obstructed blood flow of the hepatic vein and IVC, and collected the blood in the thoracic cavity for autologous transfusion. Under direct vision, the lesioned membrane was resected. The cutting end of the IVC was sutured, and the Foley's tube was removed. A non-injury vascular forceps was used to clip the cutting end of the IVC, followed by knotting of the sutures and removal the obstructed belt. If no continuous bleeding was observed, the thoracic cavity was cleansed, a pipe was used to draw liquid, and finally the cutting end was closed.

#### **Standards for therapeutic effect**

##### **Excellent**

Clinical symptoms disappeared. Ascites, superficial thoracic-epigastric varices and leg edema disappeared. The liver couldn't be felt.

##### **Improved**

Superficial thoracic-epigastric varices almost disappeared, and the liver shrank.

##### **Ineffective**

The symptoms still existed or worsened. The effectiveness included excellent and improved results.

#### **Statistical analysis**

The results were expressed by percentage, and the data were tested by the chi-square test.

## **Results**

#### **Rupture group**

In this group of 52 patients, 2 died perioperatively. Forty-four patients (85%) were followed up for 6 months to 10 years, average 6.8 years. Among them, 16 patients showed excellent results and 11 improvement with an effective rate of 61%. Seventeen patients (39%) had recurrence in 3 months (5 patients) after operation, in 6 months (7), and in 6 months (5), respectively.

#### **Interventional group**

In this group of 238 patients including 191 patients with IVC dilation and 47 patients with hepatic vein dilation, 67 had a stent inserted (58 in the IVC and 9 in the hepatic vein). Three patients died during operation. 206 (86.6%) were followed up for 6 months to 8 years, showing good or excellent results in 189 patients (91.7%), and recurrence in 17 (8.3%).

#### **Resection group**

In this group of 190 patients, 6 died perioperatively. 166 patients (87.4%) were followed up for 9 months to 8 years (average 4.9 years) with an effective rate of 90.4% (150/166) and a recurrence rate of 9.6% (16/166). In the resection group, the effective rate was 90.4% that was better than that in the rupture group ( $\chi^2=20.08$ ,  $P<0.05$ ). The recurrence rate (9.6%), however, was lower than that of the rupture group ( $\chi^2=20.08$ ,  $P<0.05$ ).

## **Discussion**

BCS is considered as retrohepatic portal hypertension and maybe accompanying with IVC hypertension.<sup>[1]</sup> Many scholars found that the commonest type of it was MOVOC in oriental countries, such as China,<sup>[1,4]</sup> Japan,<sup>[5]</sup> and India.<sup>[7,8]</sup> Though MOVOC is not as high incidence in Europe and United States, it still takes a large part.<sup>[9-11]</sup> And the treatment of MOVOC went through a long process, in 1962, Kimura<sup>[12]</sup> created the finger rupture method to treat membranous BCS through the atrium dextrum; in 1974, Eguchi<sup>[13]</sup> used the balloon dilation to treat this syndrome firstly; in 1983, Senning<sup>[14]</sup> used transcaval posterocranial resection of the liver as treatment of the BCS; in 1982, Sun<sup>[15]</sup> used the

cavoatrial shunt through post-mesenteric way and used membranotomy under extracorporeal circulation in 1986. Xu<sup>[16]</sup> applied Foley's tube to control blood flow in HV and IVC, doing membranotomy under normal temperature and direct vision. With the accumulation of knowledge about BCS and the improvement of medical technique, we found some disadvantages of this method. During the operation, the index finger of the operator needs to go a longer distance to rupture the membrane via the auricula dextran into the IVC, which limits the finger tip to rupture the membrane. After rupture and dilation of the membrane, the membrane remnants remain. When blood flows, turbulent flow may be formed, resulting in thrombosis. To avoid pulmonary embolism, this method is contraindicated when there is thrombus under the membrane. Purse suturing or clipping the auricula in an improper place may cause arrhythmia, myocardial ischemia or cardiac arrest, myocardial infarction and shock. Recurrence after rupture, large volume of blood loss and tremendous difficulty in secondary operation because of pericardium adherence, scar, and bulky vascular bypass between the lung and diaphragm result from the first operation. In some patients with scar in the IVC, the rates of rupture failure and recurrence may be higher. When the membrane is located in the IVC, it is thick and tenacious on one side and poor and brittle in the other, finger may mistakenly rupture the IVC, hepatic vein or auricule.

#### **Advantages, disadvantages and indications of interventional balloon dilation**

In 1974 Equchi<sup>[13]</sup> created interventional balloon dilation that has benefited many patients with BCS. In 1987 this technique was introduced into our hospital and has been widely used as the first choice of treatment for membranous BCS since 1990. It is mini-invasive, safe, effective, cheap and prompt, but the residual membrane and the narrowed IVC may cause occasional recurrences. In our 206 patients followed up, 17 (8.3%) showed recurrence of BCS. It was reported that complications of intervention included pericardium plugging or stent movement. Some patients with thrombus under the membrane are contraindicated for this intervention because it may induce pulmonary embolism. Some patients associated with hepatic vein obstruction should be subjected to percutaneous transhepatic dilation, otherwise nothing good result can be achieved.

#### **Advantages, disadvantages and indications of radical membrane resection**

Theoretically, membrane resection under open vision is suitable to all kinds of membranous BCS with smooth main hepatic veins or dilated hepatic veins. It is a radical treatment for direct removal of the obstruction of the IVC or the hepatic veins and reconstruction of the

normal blood stream, without change of blood direction and physiologic state. It is an ideal method to deal with membranous BCS. In our 166 patients, 150 (90.4%) had better results than those in the rupture group ( $P < 0.05$ ) and 16 (9.6%) had lower recurrence rates of BCS than those in the rupture group ( $P < 0.05$ ). Since the disadvantages of radical resection include large injury and more complications, it is unnecessary to treat all patients with this method. This method is limited to several types of BCS: intervention failures; thrombus in the IVC; recurrence after interventional therapy or often combined with thrombus in the IVC; oblique membrane in the IVC because there is tough scar around the membrane, sometimes the membrane may extend to the hepatic vein, and intervention often has high recurrence rate; foreign body in the IVC. In addition, some patients with obstruction of the IVC and hepatic veins should have the IVC and hepatic vein reconstructed together.

In summary, BCS patients should be given mini-invasive interventional treatment if they are suitable for Kimura's finger rupture. Those patients with a large lesioned membrane, thick and tough, oblique membrane or recurrence after interventional therapy should undergo radical resection. Some recurrent patients with extreme narrow or vascular defect in the IVC may receive IVC bypass out of the auricula if there is satisfied hepatic veins compensation.

#### **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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