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An Individualised Strategy and Long-Term Outcomes of Endovascular Treatment of Budd–Chiari Syndrome Complicated by Inferior Vena Cava Thrombosis

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WHAT THIS PAPER ADDS

Inferior vena cava thrombi are characterised by large individual differences and a variety of potential endovascular therapies. This study highlights a novel individualised endovascular treatment strategy based on the type, size, extent, and degree of organisation of the thrombus. The study demonstrates that this strategy can result in excellent long-term patency and favourable survival and complication rates, which is likely to have important implications for the treatment of patients with Budd–Chiari Syndrome complicated by inferior vena cava thrombosis.

Objectives: The aim was to evaluate individualised treatment and long-term outcomes of endovascular treatment of Budd–Chiari syndrome (BCS) complicated by inferior vena cava (IVC) thrombosis.

Methods: Between April 2005 and December 2015, 108 consecutive patients with BCS complicated by IVC thrombosis underwent endovascular treatment. According to the type, size, extent, and degree of organisation of the thrombus, agitation thrombolysis ($n = 7$), agitation thrombolysis combined with retrieval stent filter ($n = 5$), pre-dilation ($n = 32$), retrieval stent filter ($n = 56$), or direct large balloon dilation ($n = 8$) was performed. Peri- and post-operative follow-up data were recorded.

Results: The endovascular treatment was technically successful in 107 of the 108 patients (99.1%). The incidence of thrombosis related complications was 7.4% (8/108). Major and minor complications occurred in four patients. The mean follow-up duration was 61.7 ± 39.3 months (range 3–140 months). The cumulative 1, 2, 5, and 10 year primary patency rates were 91%, 88%, 79%, and 79%, respectively, and the cumulative 1, 2, 5, and 10 year secondary patency rates were 100%, 100%, 97%, and 97%, respectively. The cumulative 1, 5, and 10 year survival rates were 95%, 86%, and 81%, respectively. Serum albumin and total bilirubin values were independent predictors of survival.

Conclusions: For patients with BCS complicated by IVC thrombosis, an individualised endovascular treatment strategy based on the type, size, extent, and degree of organisation of the thrombus is associated with long-term patency of the IVC and favourable survival and complication rates.

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INTRODUCTION

Budd–Chiari syndrome (BCS) is a heterogeneous group of disorders characterised by obstruction of venous outflow from the level of the hepatic venules to the junction of the inferior vena cava (IVC) and right atrium of the heart.[1] The causes and types of BCS lesions vary among patients from different geographic regions[2]: in Western countries, such as Europe and the United States, obstruction of the hepatic vein by thrombosis is prevalent, whereas in Eastern countries, such as China, Japan, and Nepal, idiopathic obstruction of the IVC is the most common cause of BCS, accounting for 60%–70% of cases.[3,4] Thrombi in the IVC, which have been reported in about 20% of BCS cases, form readily because of obstruction of the IVC, stasis, turbulent or reverse blood flow in the IVC, and hypercoagulable states.[5] IVC thrombosis was once considered a contraindication to endovascular therapy because of the risk of potentially fatal pulmonary embolism from dislodgement of the thrombus after opening the blocked IVC. However, in recent years, many therapeutic approaches to treating BCS with IVC thrombosis have been advanced: anticoagulation

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therapy,[6,7] thrombolytic therapy,[8] large lumen catheter aspiration,[5,9] stent press thrombus,[10,11] agitation thrombolysis,[12] pre-dilation therapy,[13] retrieval stent filter placement,[14–16] and direct large balloon dilation.[11,17] However, optimal treatment methods for BCS with IVC thrombosis have not been defined, and the numbers of reported cases are insufficient to permit reliable assessment of long-term outcome.

IVC thromboses have been categorised as fresh thrombus, mixed thrombus, or organised thrombus. Therapy has been individualised according to the size and extent of the thrombus, and to the treatment options available, such as agitation thrombolysis, pre-dilation, retrieval stent filter, direct large balloon dilation, and combined therapy.

The purpose of this study was to establish uniform endovascular treatment strategies of BCS complicated by IVC thrombosis and to determine the long-term outcomes of BCS complicated by IVC thrombosis after endovascular treatment.

MATERIALS AND METHODS

This study was approved by the ethics committee of the First Affiliated Hospital of Zhengzhou University, and informed consent was obtained from all patients before endovascular treatment. The records of 108 consecutive patients (69 males) with BCS complicated by IVC thrombosis who underwent endovascular therapy between April 2005 and December 2015 were reviewed retrospectively. The average age was 49.4 ± 10.1 (25–74) years, and the average duration of recognised BCS disease was 72.0 ± 98.2 (0.25–480) months. Thrombi were classified as fresh, mixed, or organised, based on the findings of pre-operative imaging examinations.[5] All patients were evaluated using Doppler ultrasound, multislice computed tomography venography (CTV), and/or magnetic resonance venography, and the results were confirmed with intra-operative vena cavography. Patients with contraindications to anticoagulation ($n = 1$), severe hepatic failure ($n = 4$), or heart failure ($n = 1$) were excluded. The end of the study period was June 30, 2017. Patient characteristics are summarised in Table 1.

Table 1. Baseline characteristics of 108 patients with BCS complicated by IVC thrombosis.

Variable	Value
Sex	
M/F	69/39
Clinical manifestation	
Abdominal pain	19
Abdominal distention	62
Ascites	
Grade 1/2/3	61/9/2
Variceal bleeding	17
Hepatic encephalopathy	5
Lower extremity oedema	77
Lower extremity pigmentation	41
Site of outflow obstruction	
IVC	1
Combined HVs and IVC	107
MOVC/SOVC	90/18
Age of IVC thrombus	
Fresh	12
Mixed	88
Organised	8
Laboratory tests	
ALT (× ULN)	40.89 (4–1117)
ALP (×ULN)	93.67 (16–295)
Albumin (g/L)	47.10 (11–936)
Bilirubin (μmol/L)	30.60 (4–291.1)
Creatinine (μmol/L)	67.21 (11–338)
Blood urea nitrogen (mmol/L)	6.48 (2.2–25.7)
Prothrombin time (s)	13.89 (9.8–21.9)
Haemoglobin level (g/L)	112.63 (40–187)
Platelet count ($\times 10^9$/L)	103.17 (24–385)
Clinical prognostic indexes	
Child–Pugh score	7 (5–11)
A/B/C	37/58/13
MELD score	6.24 (−12.47–21.23)
Original Clichy score	4.61 (2.94–6.78)
Rotterdam score	0.75 (0.00–2.31)

Note. Data are medians, with ranges in parentheses. MOVC = membranous obstruction of the inferior vena cava; SOVC = segmental obstruction of the inferior vena cava; ALT = alanine aminotransferase; ALP = alkaline phosphatase; F = female; M = male; HV = hepatic vein; IVC = inferior vena cava; ULN = upper limit of normal.

Interventional procedures

All procedures were performed under local anaesthesia by two interventional radiologists with experience of 13 and 30 years in the treatment of this entity. An inferior vena cavagram was performed through a femoral approach to show the specifics of the lesion, including the IVC thrombus. Further treatment strategies and steps of endovascular treatment were directed according to the type, size, extent, and degree of thrombus organisation. The algorithm used is illustrated in Fig. 1.

The treatments of the thrombus were the following:

(a) In cases of fresh retrohepatic IVC thrombosis, agitation thrombolysis procedure was performed[12] using a pigtail catheter and guidewire to break up the thrombus. During agitation of the thrombus, a bolus of physiological saline mixed with urokinase was injected intermittently to dissolve fresh thrombus.

(b) For fresh thrombosis of the entire IVC (including the infrarenal IVC), agitation thrombolysis with retrieval stent filter (Fig. 3) was performed.[14] After agitation thrombolysis of the retrohepatic IVC, a percutaneous transluminal balloon angioplasty (PTBA) was performed with a 12–16 mm balloon catheter (Cordis, Bard, Inc., Tempe, AZ, USA) to partially dilate and achieve patency of the obstructed IVC. Then, a retrieval stent filter (Yong-Tong, Shenyang, China) was deployed, with the stent positioned across the occluded portion of the IVC (Fig. 3C).

(c) For mixed thrombosis, if the thrombus partially filled the IVC lumen (less than 75%), PTBA was performed with a 12–16 mm balloon catheter to pre-dilate the obstructed IVC (Fig. 4C) and achieve partial patency,[13] which was confirmed by contrast injection (Fig. 4D).

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