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ORIGINAL ARTICLE

Evaluation of different radiological interventional treatments of Budd–Chiari syndrome

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KEYWORDS

Budd–Chiari syndrome;
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Abstract *Aim:* To evaluate the efficacy of various interventional techniques in treatment of Budd–Chiari syndrome (BCS).

Patients methods: 103 patients with BCS were included in the study. There were 9/103 (8.7%) patients with obstruction of inferior vena cava (IVC) (type I), 17/103 (16.6%) patients with hepatic vein obstruction (type II), 71/103 (68.9%) patients with veno-occlusive diseases (type III) and 6/103 (5.8%) patients with veno-occlusive disease combined with caval thrombosis (type IV). Recanalization techniques of hepatic veins and IVC, and TIPS were used.

Results: Of all the subjects, 101 successfully underwent their procedures, with a technical success rate of 98.06%; only 2 failed to do TIPS. After treatments, 2 patients died after operation because of severe intra-abdominal hemorrhage. One hundred and one patients were followed up for 1–94 months. The mean follow-up of a BCS patient treated with PTA was 52.1 months, with an overall primary patent rate of 69.2% (18/26). The mean follow-up of BCS treated with TIPS was 33.5 months, with an overall primary patent rate of 72.7% (56/77). Eight patients died 7–64 months after the interventions.

Conclusions: Recanalization of IVC/hepatic vein and TIPS can be regarded as safe and effective interventional invasive methods in the treatment of BCS.

1. Introduction

Budd–Chiari syndrome (BCS) represents a series of pathological changes resulting from occlusive lesions in the hepatic veins and/or the inferior vena cava (IVC). The clinical manifestations of BCS include hepatomegaly, abdominal pains, ascites, and edema of the low limbs. It is an uncommon worldwide disease, but its incidence in China and other Asian countries is relatively high (1). In Western countries, primary myeloproliferative syndromes, hypercoagulable states and steroidal contraceptives were responsible for most cases. In Asian countries, pregnancy, infections and an inferior vena cava were the dominating causes (2). The prognosis for BCS is poor, and is usually difficult to cure. In recent years,

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Table 1 Classification of 103 patients with BSC based on ultrasound and angiography.

Types	*N* (%)
Type I: Membranous obstruction of the IVC W/WO thrombosis	9/103 (8.7)
Type II: Solitary hepatic vein obstruction (web/thrombosis)	17/103 (16.6)
Type III: Rudimentary (diffuse thrombosis) of hepatic veins	71/103 (68.9)
Type IV: Rudimentary (diffuse thrombosis) of hepatic veins with caval thrombosis	6/103 (5.8)

interventional therapy is widely used in the treatment of BCS as a safe and effective technique (3). However, there are still many urgent tasks not only in the selection of indicators, but also in the improvement of long-term results. Percutaneous interventional radiology procedures have been recently proposed as an alternative to surgical shunting and liver transplantation (4,5). These interventional techniques depend on the type of BCS. In 2003, a classification scheme of BCS developed and proposed type I as caval occlusion, type II as short segment (< 4 cm) isolated hepatic vein occlusion, type III as rudimentary/totally thrombotic hepatic veins and type IV that involves combined thrombosis of the hepatic veins and caval thrombosis (6).

The purpose of our study was to evaluate the feasibility and efficacy of those different interventions for the treatment of patients with BCS.

2. Subjects and methods

This retrospective cohort study was conducted at the National Liver Institute, Menoufiya University from October 2006 to April 2013. It included 103 patients having Budd–Chiari syndrome who were selected and submitted for the treatment with interventional procedures. All patients had informed consents before interventions. Generally, there were 73 females and 30 males. Their ages ranged from 14 to 44 years, with a mean of 29.5 years. The duration of symptoms ranged from 1 to 10 years. Clinical symptoms observed included abdominal pain and distention, hepatomegaly, splenomegaly, ascites and varicose veins in the abdomen and legs.

On the basis of angiography and ultrasonography, 103 patients were classified into 4 types of BCS (Table 1): type I solitary obstruction of the IVC by membrane/thrombosis (9/103; 8.7%), type II solitary short segment/membranous hepatic vein obstruction (17/103; 16.6%), type III rudimentary/diffuse thrombotic obstruction of the hepatic veins (veno-occlusive disease) (71/103; 68.9%) and type IV combined thrombosis of the hepatic veins and retro-hepatic cava (6/103; 5.8%). For those of caval obstruction, the length of obstructed IVC ranged from 1 to 2 cm and the width ranged from 16 to 22 cm. Obstructive hepatic vein ranged from 1 to 4 cm and 0.8 to 1 cm wide. All operations were performed under the guidance of standard angiography (Fluoroscopic X-ray DIGITAL unit: infinix, Toshiba, Japan) with ultrasound guidance (Toshiba-xario with 5 MHZ convex transducer and Toshiba nemio XG with 3.75 MHZ convex transducer).

2.1. Methodology

2.1.1. Treatment of type I Budd–Chiari by caval recanalization (9 patients)

Caval recanalization by dilatation with or without shunting is considered the best option for such patients. In all patients, we used primarily the femoral vein to obtain inferior vena cavography to evaluate for the site and degree of occlusion. In cases of incomplete occlusions, the contrast passed to opacify the heart. In such cases, we passed the guide wire through the stenosis followed by the large-diameter balloon (20–24 mm) and dilatation was obtained. If occlusion was complete, RUPS-100 (Rosch-Uchida Puncture Set) device or J-type Brockenbrough needle (Cook, Chicago, USA) was passed from the jugular vein. While the pigtail catheter was in cava (from femoral vein), lateral view was obtained to have alignment straight positioning. Once optimal alignment was obtained in anterior and lateral projections, the Rups100 or Brockenbrough needle was pushed slowly to penetrate the occlusion segment till it reaches the other side. Blood was aspirated to confirm luminal position and then wire was advanced while the needle is removed. The large-diameter balloon was then advanced to dilate the segment (Figs. 1 and 2).

2.1.2. Treatment of type 2 by hepatic vein recanalization (17 patients)

For recanalization of one of the hepatic veins, transjugular or trans-hepatic approach was used. In some cases, combined approaches were used. As a rule, we used the transjugular approach at first, where a long sheath was inserted. Catheter manipulation was done to go through the narrowed ostium. After passing, 10–12 mm balloon was inserted and the ostium was dilated. In case of failure to pass through the stenosis (total occlusion), the RUPS-100 needle was inserted at the osteal occlusion and pushed to get the balloon after. If transjugular approach failed, trans-hepatic approach was used. Under US guidance, the proper hepatic vein was punctured and manipulation was performed to pass the occluded segment to get the dilatation (Figs. 3–5).

2.1.2.1. Shunting of cava and hepatic vein. We used shunts for patients with type 1 and type 2 in case of immediate recoil at the time of operation, in case of failure to dilate or in late cases with recurrent stenosis/occlusion.

For caval shunting, balloon-mounted shunts of diameter 2–4 mm larger than the diameter of cava and longer by 2–3 cm from that of the occlusion length. The shunt was introduced through the suitable sheath (12 Fr). The shunt set was advanced