Interventional endovascular treatment of Budd-Chiari syndrome with long-term follow-up

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Purpose: To present results of a 9 year follow-up of percutaneous transluminal angioplasty (PTA) and stents placement in patients with Budd-Chiari Syndrome (BCS) and to evaluate the clinical value and applicability of this approach.

Materials and methods: 44 consecutive patients with BCS (25 male and 19 female; average age, 42.6 years; age range, 19–77 years) were treated with PTA and stents during a 9-year period. The mean duration of symptoms was 46 months. Underlying active malignancy was the cause of occlusion in 5 patients. 3 patients had a history of taking oral contraceptives. The obstructed inferior vena cava (IVC) or hepatic vein (HV) were first dilated by a percutaneous transluminal balloon, and then a self-expanding stent was placed. Clinical patency was defined as absence or improvement of symptoms. Clinical follow-up was supplemented with colour Doppler sonography, CT scan, or both.

Results: Technical success was achieved in 97.2% (35/36) IVC and 83% (10/12) HV PTA and stents. Significantly, the IVC pressure decreased from 2.7 kPa (SD = 0.3) to 1.5 kPa (SD = 0.4) and HV pressure dropped from 2.3 kPa (SD = 0.4) to 1.3 kPa (SD = 0.2). The symptoms and signs disappeared or were relieved after operation in most of the patients. A few serious related complications including one stent migration and one pulmonary emboli occurred and were resolved in time. Patients were followed for a mean of 44 months (range 3–102). Short- and long-term results were satisfactory except for 3 patients (9.4%, 1 IVC, 2 HV) who presented with a restenosis or thrombosis and underwent additional therapy. There were 5 deaths owing to underlying malignant disease 3–17 months after the procedures.

Conclusion: PTA and stent placements proved a safe and effective treatment in BCS and had a good long-term outcome and should be considered in patients who have symptoms or have no adequate alternative therapy.

Key words: Budd-Chiari syndrome; percutaneous transluminal angioplasty; stent

Introduction

Budd-Chiari syndrome (BCS) is a rare disease characterised by obstruction of outflow in the hepatic vein (HV) and/or the inferior vena cava (IVC). It often occurs secondary to intrinsic vascular thrombosis, hepatic tumour invasion/compression, or is associated with an idiopathic obstructed membrane leading to hepatomegaly, portal hypertension, impaired liver function, formation of communicating channel, and oedema or ulcer in lower extremities [1, 2]. The most common type in Asia is short length obstruction (membranous and segmental) in the IVC and/or in the ostium of the main HV. Most of them are chronic and idiopathic. In Western countries thrombotic obstruction is the most common cause [3]. The clinical signs of ascites, abdominal pain and hepatomegaly are the typical triad of BCS. Since these signs are non-specific, the clinical diagnosis of this syndrome is difficult. At present, colour Doppler sonography (CDS), contrast-enhanced computed tomography (CCT) and magnetic resonance imaging (MRI) are the effective non-invasive imaging techniques used in the evaluation of the patency of the HV and the IVC.

In the last decade, surgical therapy proved to be effective for some patients but was known to be associated with a considerable morbidity and mortality, especially in some weak cases [4, 5]. Surgery
was contraindicated in some cases because of the patient’s elderly age and the concomitance of associated pathologies. Percutaneous transluminal angioplasty (PTA) and stent placement were important innovations for the treatment of central venous disease. Due to its minimal-invasive approach, endovascular procedure was applicable for patients with contraindication to major surgery. Nowadays it has become a valid alternative for the major portosystemic shunts with beneficial results [6, 7]. Up to now, most of the reports were based on relative small numbers of patients, with short- and intermediate-term relief of such large venous obstructions. PTA and stent placement of the HV or the HV and the IVC simultaneously were limited [8]. The overall endovascular management of BCS also needed more exploration.

Materials and methods

From April 1995 to June 2004, 44 patients with BCS underwent stent placement in our hospital. There were 25 males and 19 females. The average age was 42.6 ± 6.3 years (range 19–77 years). The mean duration of symptoms was 46 months, ranging from 2 months to 22 years. Before operation, the diagnosis of BCS was confirmed by clinical examination: all patients underwent colour Doppler sonography, 12 patients underwent CCT scan and 8 patients underwent venography. Most of the etiological factors were unclear. Underlying active malignancy was the cause of occlusion in 5 patients, including 4 hepatocellular carcinoma (HCC) and 1 suprarenal tumour. In 2 patients, IVC obstruction was due to superior extension of pelvic thrombophlebitis. 3 patients had a history of taking oral contraceptives. Patients had chronic symptoms and signs of IVC and HV occlusion. 44 patients were divided into obstruction of IVC with at least one patent HV (IVC group, n = 32 patients), obstruction of three main hepatic veins (HV group, n = 8 patients), obstruction of both IVC and three main hepatic veins (combined group, n = 4 patients). Of the 36 patients who underwent IVC stent placement, 21 patients had membranous obstruction, and 15 patients had IVC segmental obstruction (range 1.1–5.5 cm). IVC stent were placed in both the IVC group and the combined group (36 patients). All membranous lesions in IVC were suprahepatic IVC obstruction. While in 12 patients who underwent HV stent placement, 8 patients had membranous obstruction in the HV and 4 patients had segmental obstruction in the HV (range 1–2 cm). HV stent placement was performed in both the HV group and the combined group (12 patients). Furthermore, patients with long segmental obstruction due to thrombosis (more than 6 cm) were excluded from this study (table 1–2).

The interventional methods were approved by the ethic committee of our hospital. The risks, benefits, and alternatives of the procedure were explained to all patients, and written informed consent was obtained.

Endovascular procedure

All the procedures were performed in an angiography-suite, with stand-by for cardiopulmonary bypass.

<table>
<thead>
<tr>
<th>Occlusive site</th>
<th>IVC (n = 32)</th>
<th>HV (n = 8)</th>
<th>combined (n = 4)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Signs</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>ascites</td>
<td>22</td>
<td>6</td>
<td>4</td>
</tr>
<tr>
<td>distended abdominal veins</td>
<td>29</td>
<td>5</td>
<td>4</td>
</tr>
<tr>
<td>hepatomegaly</td>
<td>17</td>
<td>8</td>
<td>4</td>
</tr>
<tr>
<td>splenomegaly</td>
<td>26</td>
<td>6</td>
<td>3</td>
</tr>
<tr>
<td>leg oedema</td>
<td>30</td>
<td>7</td>
<td>4</td>
</tr>
<tr>
<td>leg ulcer</td>
<td>15</td>
<td>3</td>
<td>3</td>
</tr>
<tr>
<td>Symptoms</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>weakness</td>
<td>30</td>
<td>8</td>
<td>4</td>
</tr>
<tr>
<td>abdominal fullness</td>
<td>32</td>
<td>8</td>
<td>4</td>
</tr>
<tr>
<td>gastrointestinal bleeding</td>
<td>11</td>
<td>3</td>
<td>2</td>
</tr>
<tr>
<td>jaundice</td>
<td>5</td>
<td>3</td>
<td>2</td>
</tr>
<tr>
<td>hepatic encephalopathy</td>
<td>2</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>low extremities pain and cramps</td>
<td>7</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>decreased exercise tolerance</td>
<td>11</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Others</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>anaemia</td>
<td>15</td>
<td>3</td>
<td>2</td>
</tr>
<tr>
<td>hypoproteinaemia</td>
<td>20</td>
<td>5</td>
<td>4</td>
</tr>
<tr>
<td>renal failure</td>
<td>12</td>
<td>3</td>
<td>2</td>
</tr>
</tbody>
</table>
Patients were placed in supine decubitus and the invasive venous pressure, electrocardiogramme and urine output were monitored. All of the procedures were carried out under local anaesthesia and no cases were necessary to convert to other anesthesia during the procedures. The intervention was performed in the operating room by a team consisting of a vascular surgeons and an interventional angiologist. Before the procedure, all the patients were given Aspirin 300 mg and Livzonphin 1.0 unit daily for 3 days. In 5 patients with massive ascites, before the procedure a therapeutic paracentesis was performed followed by intravenous administration of albumin. In some patients, the cardiogenic or hepatic malfunction were improved by correlative drugs before operation.

First, a 5F sheath was advanced through a percutaneous right femoral vein access, after heparin was administered intravenously (1 mg/kg body weight), a pigtail catheter was inserted over a 0.035 guide wire into the IVC, and intermittent contrast angiography was performed to confirm the obstruction situation and to evaluate the feasibility of endovascular therapy. If the obstruction was incompletely, the soft end of a guide wire could easily cross the narrowed part of IVC into the right atrium. For complete obstruction, the hard end of the guide wire was pushed carefully into the occluded IVC and went straight through the lesion. If necessary, a Brockenbrough needle was inserted to cut through the lesion. Once the obstruction was broken through, the needle of the guide wire was stopped immediately and was exchanged by a 4F catheter and intraoperative angiography was performed immediately. If the end of the catheter was confirmed to have been placed into the right atrium, then the Amplatz ultra-stiff (Boston Scientific Corporation, Miami, FL, USA) guide wire was exchanged and, depending on the lesion situation, a balloon catheter with a 1.6–2.6 cm diameter was inserted to dilate the IVC 2–3 times. Second angiography was performed to evaluate the dilation effect. Then, through the Amplatz ultra-stiff guide wire, a 12F sheath (COOK, USA) containing the stent (Gianturco stent, Zhiye Medical Equipment, Changzhou, China) was pushed into the obstructed part of the IVC and the stent was deployed to support the obstructed portion. The progression was controlled by radioscopy, and the central venous pressure (CVP) and ECG were examined. The choice of the diameter and length of the stent were based on an analysis of pre-operative examinations, where a stent 20% bigger than the diameter of the normal IVC was always used. At the end, the last angiography was performed to confirm the stent’s position and effect. If the stent was not expanded completely, it was necessary to blow up carefully the balloon so as to better anchor the stent to the IVC (most stents could expand itself). The venous pressure of IVC was measured before and after endovascular treatment.

If the obstruction of the HV was found, a 5F sheath was advanced through a percutaneous right femoral vein access, after heparin was administered intravenously (1 mg/kg body weight), a pigtail catheter was inserted over a 0.035 guide wire into the IVC, and intermittent contrast angiography was performed to confirm the obstruction situation and to evaluate the feasibility of endovascular therapy. If the obstruction was incompletely, the soft end of a guide wire could easily cross the narrowed part of IVC into the right atrium. For complete obstruction, the hard end of the guide wire was pushed carefully into the occluded IVC and went straight through the lesion. If necessary, a Brockenbrough needle was inserted to cut through the lesion. Once the obstruction was broken through, the needle of the guide wire was stopped immediately and was exchanged by a 4F catheter and intraoperative angiography was performed immediately. If the end of the catheter was confirmed to have been placed into the right atrium, then the Amplatz ultra-stiff (Boston Scientific Corporation, Miami, FL, USA) guide wire was exchanged and, depending on the lesion situation, a balloon catheter with a 1.6–2.6 cm diameter was inserted to dilate the IVC 2–3 times. Second angiography was performed to evaluate the dilation effect. Then, through the Amplatz ultra-stiff guide wire, a 12F sheath (COOK, USA) containing the stent (Gianturco stent, Zhiye Medical Equipment, Changzhou, China) was pushed into the obstructed part of the IVC and the stent was deployed to support the obstructed portion. The progression was controlled by radioscopy, and the central venous pressure (CVP) and ECG were examined. The choice of the diameter and length of the stent were based on an analysis of pre-operative examinations, where a stent 20% bigger than the diameter of the normal IVC was always used. At the end, the last angiography was performed to confirm the stent’s position and effect. If the stent was not expanded completely, it was necessary to blow up carefully the balloon so as to better anchor the stent to the IVC (most stents could expand itself). The venous pressure of IVC was measured before and after endovascular treatment.

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The average operation time was 45 minutes (range from 30 to 85 minutes). After the end of the procedure and angiographic check, the systems and guides were removed. The macasirol (20 mg) and cedilanid (0.2 mg) were administered intravenously to prevent acute congestive heart failure and the urine volume was monitored for two days. Antibiotics were given to all patients for two days. All patients underwent anticoagulation therapy with heparin sodium during the procedure and received warfarin sodium (Coumadin; Dupont, Wilmington, Del) to achieve an international normalised ratio, or INR, of 2–3 for at least 6–8 months. Longer anticoagulation therapy was instituted for patients with uncorrectable underlying hypercoagulable states.
Results

The result of the procedure was considered to be successful when the pressure gradient disappeared after the balloon dilation and stents deployment in the correct position. All patients underwent colour Doppler scan to confirm the stents status before discharge. Thereafter, they were seen at 3–6 month intervals, or when they had recurrence of symptoms of BCS.

An IVC stent was placed successfully in 97.2% (35/36 patients) of the patients (IVC and combined groups). The procedure failed in one patient with a segmental occluded IVC (5.5 cm), and the Brockenbrough needle failed to cut through the hard occlusive segment of the IVC. Pulmonary emboli in one patient and arrhythmia in 2 patients occurred during balloon dilation. One stent migrated into the right atrium in one patient and was removed in a later radical operation. No procedural death occurred. Haemodynamic features in patients with successful stent placement improved significantly, the inferior vena cava pressure below the obstruction decreased from 2.7 kPa (SD = 0.3) to 1.5 kPa (SD = 0.4), and satisfactory antegrade flow in the IVC was observed in follow-up with a normal flow spectrum in CDS (table 3). All patients in the IVC group improved clinically. Ascites, hepatosplenomegaly, lower extremity oedema, and distended abdominal veins disappeared or diminished obviously at discharge.

26 of 31 patients in the IVC stent group were followed for a mean of 54 months (3–102 months). The other 5 patients were lost for follow-up or died from underlying malignant disease. Of the remaining 26 patients, 21 had no symptoms and 5 improved clinically but had still mild weakness or abdominal swelling on physical examination. Of the 4 patients in combined group, 1 was lost to follow-up and the other 3 patients had no symptoms of IVC obstruction. All of the followed patients (29/35 patients) in the IVC and combined group were examined by CDS and showed that the IVC stent was patent and worked effectively in 28 patients. The overall IVC stent patency rate was 96.6% (28/29 patients). Stent occlusion occurred because of compression in one patient at 6 months following stent placement, and had recurrence of abdominal fullness, oedema of lower extremities and large quantity of ascites. This patient was treated again with balloon catheter dilation, diuretics and anticoagulants and the 3 years follow-up showed a patent stent in this patient. Two stents that were compressed in the procedure expanded themselves gradually within the two weeks after the operation.

In 8 patients of the HV group, 6 stents were placed successfully alone, although some patients had narrowed IVC pressed by enlarged caudate lobe. In 4 patients of the combined group, successful IVC stent placement resulted in disappearance of all symptoms of IVC obstruction. However, ascites, and hepatosplenomegaly were not alleviated. Therefore, a hepatic vein stent was placed 1 week after the IVC stent placement. Together, in the patients of the HV and the combined group, all HV stents were successfully placed in 83.3% (10/12 pa-
tients) of the patients. In 2 patients of the HV group the operation failed due to a long occluded hepatic vein (2.5 and 3.0 cm), which was difficult to cut through. All of the 10 successful patients had haemodynamic improvement immediately after the procedure. HV pressure dropped from 2.3 kPa (SD = 0.4) to 1.3 kPa (SD = 0.2) and satisfactory contrast flow was noted in the stented HV. At the time of discharge, 5 patients were free of ascites, the massive ascites decreased without diuretics, and hepatomegaly disappeared or diminished obviously in 8 of 10 patients.

Early stent occlusions occurred in 2 patients of the HV group on the second day after the procedure, because of one stent thrombosis and another stent migration into the distal hepatic vein. The first one refused endovascular therapy and the other one accepted the second HV stent. 2 patients with peritonitis and ascites were treated with macasirool and parenteral antibiotics. One patients experienced pericardial effusion which was resolved within one week. No other major complications occurred.

There were 9 patient (6 in the HV group, 3 in the combined group) who were followed during a period of 43 months (range 6–71 months). Clinical symptoms including ascites, hepatomegaly and splenomegaly were greatly alleviated or disappeared. The rest of the patients were lost for follow-up or died from underlying malignant disease. Periodic liver function tests were consistently normal or improved. CDS examinations demonstrated that the overall HV stent patency rate was 77.8% (7/9 patients). One young woman was pregnant and delivered a healthy baby. Two new HV stent occlusions were treated with drug therapy as the patients did not agree to be operated upon (table 4–5).

### Table 5

The results of follow-up patients.

<table>
<thead>
<tr>
<th>IVC obstruction (n = 32)</th>
<th>HV obstruction (n = 8)</th>
<th>IVC and HV obstruction (n = 4)</th>
</tr>
</thead>
<tbody>
<tr>
<td>successful stenting (n = 31)</td>
<td>successful stenting (n = 6)</td>
<td>successful stenting (n = 4)</td>
</tr>
<tr>
<td>followed (n = 26)</td>
<td>followed (n = 6)</td>
<td>followed (n = 3)</td>
</tr>
<tr>
<td>patency (n = 25)</td>
<td>occlusion (n = 1)</td>
<td>patency (n = 3)</td>
</tr>
<tr>
<td>occlusion (n = 1)</td>
<td>occlusion rate</td>
<td>occlusion (n = 0)</td>
</tr>
</tbody>
</table>

**Discussion**

BCS refers to a collection of disorders. In the US and Europe this syndrome is considered to include hepatic venous occlusive disease, whereas it is more commonly considered to include a suprahepatic inferior vena cava occlusive lesion in South Africa and eastern Asian countries [9, 10]. We define BCS as a rare type of portal hypertension caused by complete or incomplete obstruction of the hepatic veins or the suprahepatic IVC, or both. It is characterised by the clinical manifestations of portal hypertension either with or without IVC hypertension. With increasing awareness of this disease and the development of medical imaging technology, more and more cases have been reported in many countries. Early diagnosis is achieved by CDS or CCT scan, the latter giving a typical image with stagnation of contrast material. Cavography is highly recommended to select the best procedure and to identify possible involvement of the vena cava [9].

BCS is more common in East Asia than in Western countries. The majority of patients are found in China, India and Japan. The prevalence in China is reported to be 0.0065%, being the highest along the lower bank of the Yellow River and in three target counties [10]. The aetiology of
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BCS is not yet elucidated. Next to the congenital forms, there are forms associated with tumours, polycythaemia vera and myelodysplastic syndrome, thrombophilia including factor V Leiden and the antiphospholipid syndrome, oral contraceptives, paroxysmal nocturnal haemoglobinuria, granulomatous idiopathic venulitis and hypersesinophilic syndrome [11–17].

The natural history of BCS is not completely known. However, it is clear that the majority of untreated patients die from the sequelae of this disease. Of the 34 untreated cases only 2 markedly improved the symptoms during 6 years of observation. 18 of the untreated cases died within 7 years. The prognosis of BCS depends on such factors as cardiac function, liver damage, extension of thrombosis, recanalisation and compensatory anastomoses. Death from complication of haematemesis from oesophageal varices and liver cancer were the most frequent, followed by hepatic coma, deterioration of the general condition by protracted ascites and malnutrition. Early mortality is high – up to 30%; the presence of oesophageal varices was a poor prognostic sign [18].

The treatment depends mainly on the mode of presentation, the presence of liver failure and anatomical considerations. The management of BCS traditionally consisted of conservative treatment, portal decompression, hepatopetal anastomosis, transjugular intrahepatic portosystemic shunts (TIPS), endovascular thrombolysis, PTA and stents. Liver transplantation as the second surgical option for BCS was indicated for BCS in acute or chronic liver failure in the Western world [19, 20]. Conventional medical therapy with diuretics and anticoagulation has been reported to be of limited value in relieving hepatic venous outflow obstruction and TIPS usually lead to hepatic coma [21]. In the last decade, the surgery, which included cavoatrial shunt, mesoatrial shunt, splenoatrial shunt, cavocaval shunt, transcatheter membranotomy and liver transplantation, has been shown to be effective for selected patients with BCS [22].

Sometimes surgery was contraindicated in some cases because of the patient's weakness and the concomitance of associated pathologies. Venous bypass surgery has been the only option in some selective severe cases and has had limited efficacy and applicability [23, 24].

The treatment of BCS using endovascular stents was an important innovation for these diseases. Due to its minimal-invasive approach, endovascular stenting was also applicable for patients with contraindication to major surgery. It has now become a valid alternative to medical and traditional surgical therapy. It remarkably decreased the risk, and it seemed to improve the survival and quality of life of these patients. In our series, BCS was caused by short-medium length (less than 6 cm) obstruction of the hepatic IVC and/or of the main HV. Most patients were chronic and idiopathic, and stent placement was performed based on the locations of vascular obstruction. Our experience showed that CDS could identify the site, degree and extent of obstruction of hepatic IVC or hepatic veins and demonstrate that the altered haemodynamic within the IVC and the HV, was the most convenient shortcut to screen our patients. However, with respect to the patients with malignancy, CCT, MRI or cavography were recommended to confirm the exact occluded site and to evaluate the feasibility of endovascular therapy.

IVC obstruction with membrane or short length lesion is a common cause for BCS in the Eastern countries [1, 25]. Endovascular balloon dilation has been proven to be effective in a great majority of patients with stenosis or occlusions of major veins. It is performed to reestablish venous flow and to relieve symptomatic venous obstructions secondary to benign disease, malignant disease, and/or radiotherapy and has been associated with little morbidity and mortality. In the early study 10 years ago, we treated some patients only with balloon angioplasty. However, in most cases the residual membrane was often pushed back into the lumen of the IVC once the balloon catheter moved back. The restenosis incidence was as high as 75% (9/12 cases). Almost half of them had to accept redilation, even in patients with membranous obstruction [26]. It seemed that the results of angioplasty alone were also disappointing, particularly in the setting of chronic venous occlusions due to venous recoil, low flow state, and thrombogenicity of the lumen [27, 28]. Ever since stents were placed in the restricted part in all patients, irrespective whether the lesion was a membranous or a segmental obstruction, the restenosis incidence dropped to 7% (2/29). Based on our follow-up data, IVC stent placement showed to be effective and reliable in management of BCS and had a satisfactory long-term patency.

Concerning the complete obstruction of IVC with hard membrane or short length lesion, the direction of the Brockenbrough needle was difficult to adjust and it was possible that pericardial effusion occurred when the heart was hurt by the needle. It was very dangerous. However, when the procedures were divided into 2 consecutive steps, the treatment became simple. First, through a percutaneous femoral vein approach, the primary cavography was performed. If the contrast medium could not flow into atrium, a pigtail catheter was inserted over a guide wire into the right atrium through a percutaneous right cervical vein access, and intraoperative angiography was performed to confirm the distance and direction between the retro bottom of the right atrium and the obstructed top of the IVC. In this way, the feasibility of endovascular therapy could be assessed. The sharp instruments, probed with the back end of a wire, were required to connect the short segments between the superiorly and inferiorly created channels. The wire introduced through the jugular access was then snared from the groin by using a vascular snare (Amplatz; Microvena, St Paul, Minn) to allow femoral access to the entire length of the oc-
cluded IVC. The occluded venous segments were then predilated by using a balloon of smaller calibre than the IVC to allow stent placement. This method was used successfully in eight patients of our series with IVC segmental obstruction. If the difficulty was met in access, the right cephalic vein or basilic vein could be available.

Due to the difficulty in locating the ostium of the HV and its small diameter, the treatment of HV obstruction was a challenge in the treatment of BCS [1, 29]. It was very difficult to cut through the occluded orifice of the hepatic vein from the vena cava. We performed percutaneous hepatic vein recanalisation, dilation and stent placement through the percutaneous right cervical or femoral vein access and HV stents were successfully placed in 83.3% (10/12) patients. However, according to our follow-up data, the later patency rate of the HV was lower than that of the IVC. We estimated that, during a long-term follow-up, almost one third of the patients suffered from late stent occlusion including several cases that were lost for follow-up. We considered that venous recoil, low flow state, thrombogenicity and impairment of the vein wall during the procedure were the main factors leading to later stent obstruction. Due to limited cases, we could not assess exactly the different patency rate between the HV group and the combined group. At present, we are studying a new type wall stent and a delivery system of the HV procedure to improve the patency rate.

With regard to the management of the combined IVC and HV obstruction, based on our clinical experience, the two steps procedure was also recommended due to the difficult operation [30]. After the first failed HV stenting in one of the patients, all of the four stents were placed two weeks after the IVC stenting. The patent IVC and completely expanded IVC stent helped to place the second stent and, in the same time, the necessity for it could be reevaluated. During a long-term follow-up, only one patient developed HV stent occlusion. The other IVC stents worked well during the long-term follow-up. Based on our operative experience, it was necessary to dilate the HV stent after the stent placement to prevent possible compression from the IVC stent. The long-term follow-up results showed that the combined procedure was an acceptable choice for BCS patients with both IVC and HV obstruction.

With regard to late stent occlusion, based on our statistical data, absence of long-term anticoagulants was the important factor related to stent occlusion. In our series 2 out of 3 occlusive stent cases could not keep anticoagulant therapy. Therefore, anticoagulants are strongly recommended for about 12 months after the procedure, especially in the HV stent cases.

Based on our experience, the three segments stent proved to be more suitable than the two segments stent in an endovascular procedure for the exact localisation. Avoiding the stent migration, the stent junction between the first and the second segment must be delivered exactly against the stenosis site of the IVC. Due to the characters of Giantureco Z stents, awareness of stent migration is in place. If the stent could not expand completely during the procedure, the balloon was recommended to be blown up after a few days or weeks, because the stent could usually expand itself gradually. As a serious accident, one IVC stent slid into the right atrium during the balloon dilation and was removed in an emergent radical operation.

In our past series of some patients with symptoms of chronic IVC occlusion or terminal malignancy, supportive measures were frequently the only therapy offered on account of the invasive procedure and associated morbidity of the surgical alternatives. In our series, the endoluminal approach was effective in relieving the symptoms in these patients. We consider this active therapy the first line of treatment. Given the patients’ short life expectancy its goal is mainly palliative. In 5 of our patients with occluded IVC due to unresectable underlying malignancy, the symptoms of Ascites and leg swelling reappeared rapidly after the successful IVC stenting.

A few complications were encountered in our series of patients, which calls the attention to the safety of this approach. We encountered two serious complications: one acute pulmonary emboli after initial balloon dilation and one stent migrating into the atrium as described above. To avoid pulmonary emboli, for the two later thrombosis of IVC, we inserted the guide wire through the thrombosis and delivered the stent directly to cover the whole thrombosis site. The thrombus shedding did not occur without balloon dilation. In addition, four patients had pain that resolved spontaneously without any sequelae during balloon dilation and stent placement in the IVC.

In addition to the interventional procedures, 12 other patients in our hospital underwent surgical procedures including membranotomy (3 cases), mesoatrial shunt (7 cases), radical resection (2 cases). 2 patients underwent mesoatrial shunt and 1 patient who underwent radical resection died of encephalopathy or respiratory infection. For the other patients, the symptoms and signs disappeared or were relieved after operation. With respect to the indication for surgery or PTA, in our opinion, all of the membranous obstructions and short segmental obstructions (range 1.1–5.5 cm) are recommended to undergo the interventional procedure. On the other hand, serious liver function failure, weakness and advanced age are also indications for PTA plus stent deployment based on their high surgical risk. However, for patients with long segmental obstruction (more than 6 cm), IVC thrombosis, solid or inclined membranous obstruction surgical procedures are recommended.

Furthermore, we consider that stenting should not be performed in patients with recurrent thrombosis and restenosis after PTA plus stenting. In this case, surgical procedures such as radical cor-
Interventional endovascular treatment of Budd-Chiari syndrome with long-term follow-up


References


13. Pelletier S, Landi B, Piette JC, et al. Antiphospholipid syndrome in paroxysmal nocturnal hemoglobinuria. A spectrum from venous occlusive disease to venous outflow obstruction in patients with idiopathic BCS when the underlying lesion is not amenable to angioplasty only. Furthermore, more studies are necessary to determine the role of stenting in the treatment of HV and long segmental IV of obstruction. We suggest that endovascular stenting may prove to be a valid therapeutic alternative for surgery for certain forms of central venous diseases.

Correction or liver transplantation are required. Certainly, the prior stenting will lead to technical difficulty in operations on veins. The suprarenal IVC part must be resected with a stent if necessary. In a radical operation, the stent must be removed from the IVC, to prevent recurring thrombosis. However, we felt sorry to be short of correlation experience. Of course, it would not affect surgical procedures such as cavoatrial shunt, mesoatrial shunt, splenoatrial shunt or cavocaval shunt.

Together, with a high successful rate during almost 9 years of long-term follow-up, IVC and HV stents or combined IVC and HV stents could be applied to certain BCS patients depending on the sites of obstruction. The dilation and stenting plus additional surgery proved a prosperous means for treating BCS with localised lesions. Primary stent placement could serve as the first line of treatment in patients with idiopathic BCS when the underlying lesion is not amenable to angioplasty only. Furthermore, more studies are necessary to determine the role of stenting in the treatment of HV and long segmental IVC of obstruction. We suggest that endovascular stenting may prove to be a valid therapeutic alternative for surgery for certain forms of central venous diseases.
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