

Non-TIPSS Management of Budd Chiari Syndrome

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Abstract

Background: Budd Chiari syndrome (BCS) is a heterogeneous group of vascular hepatic disorders. Imaging with recent advances in these techniques play an important role in diagnosis and management of cases with BCS. The aim of the current work was to study the clinical effectiveness of venoplasty, without the need of performing a TIPSS procedure, in management of BCS.

Aim of Study: The aim of the current work was to study the clinical effectiveness of venoplasty, without the need of performing a TIPSS procedure, in management of BCS.

Patients and Methods: Clinically, all patients presented with symptoms of abdominal pain and distention and signs of hepatomegaly and ascites.

Results: Clinically, all BCS patients presented with symptoms of abdominal pain and distention and signs of hepatomegaly and ascites were included. Informed consent was taken from all patients. These cases were managed as follows: Three cases: IVC angioplasty. Two cases: IVC angioplasty and stent. Four cases: Hepatic vein angioplasty. Four cases: Hepatic vein angioplasty and stent. One case: Collateral hepatic vein angioplasty.

Conclusion: Budd Chiari patients should have a thorough Doppler examination to assess the presence of a communication between the hepatic vein or their intrahepatic collaterals with the IVC. The presence of this venous communication could allow us to perform a venoplasty with/without stenting and avoid performing unnecessary TIPSS procedure. Venoplasty has a very good long-term outcome in Budd Chiari syndrome.

Key Words: Budd Chiari syndrome – TIPSS – Stent – Angioplasty – Venoplasty – Collaterals – IVC – Hepatic veins.

Introduction

Budd Chiari syndrome (BCS) is a heterogeneous group of vascular hepatic disorders that is characterized by occlusion of one or more of the hepatic veins (HV) or occlusion at the level of hepatic

portion of inferior vena cava (IVC) with consequent development of portal hypertension [1-4].

BCS can be classified according to the etiology to primary and secondary, according to level of obstruction: Type I (obstruction of IVC with or without HV) II (hepatic veins obstruction) and III (small centrilobular venules obstruction) or according to disease duration (acute and chronic) [5].

Clinical presentation is variable, it may be asymptomatic or minimal complain up to portal hypertension, ascites, hepatomegaly, splenomegaly, variceal bleeding and even liver failure. In acute type hepatomegaly with thrombosed HV and ascites are common. In chronic cases there is membranous or fibrotic obstruction of HV and/or IVC [1].

Imaging with recent advances in these techniques play an important role in diagnosis of cases with BCS including Doppler ultrasound for assessment of patency of IVC and hepatic veins, CT and MRI to diagnose not only the level of venous occlusion but also altered texture of the liver with development of nodular changes and collaterals. Features are different according to the stage and duration of the disease [2,6].

Different management techniques for BCS are present including transjugular intrahepatic portosystemic shunt (TIPSS), surgical shunts, recanalized occluded hepatic veins by angioplasty with stent and even liver transplantation. Balloon angioplasty may be difficult in long standing cases with long fibrotic occluded venous segment [7].

List of Abbreviations:

TIPSS : Transjugular intrahepatic portosystemic shunt.
BCS : Budd Chiari syndrome.
HV : Hepatic veins.
IVC : Inferior vena cava.
CT : Computed tomography.
MRI : Magnetic resonance imaging.

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Patients and Methods

Abdominal and chest wall varicosities were found in four patients.

This is a retrospective study of Budd Chiari syndrome cases referred to Radiology Department, Alexandria University for interventional management in the period from September 2016 till August 2018.

Inclusion criteria:

- Patients diagnosed with BCS on clinical and imaging basis including CT scan and Doppler examination.
- Presence of at least one patent intra-hepatic vein in continuation with the IVC.

Exclusion criteria:

- Patients with impaired renal function.
- Patients with coagulopathy (INR >1.5) or thrombocytopenia (platelets <50,000).
- Complete IVC and/or all hepatic veins occlusion.

Procedures performed for management of BCS in the study:

- Three cases: IVC angioplasty.
- Two cases: IVC angioplasty and stent.
- Four cases: Hepatic vein angioplasty.
- Four cases: Hepatic vein angioplasty and stent.
- One case: Collateral hepatic vein angioplasty.

Details of the procedures:

Informed consent was taken from all patients.

All procedures were done under local anesthesia and intravenous analgesics and complete aseptic technique. All venous punctures were done under ultrasound guidance using Toshiba Xario (5 MHz convex transducer).

All patients are kept on anticoagulant therapy for life with monitoring of the INR between 2 and 2.5.

I- IVC Angioplasty and stent:

This procedure was done in five patients where the hepatic IVC showed a focal stenosis. In three cases, this was the definitive treatment. While in two cases, an IVC stent was placed.

Antegrade puncture of the right femoral vein was done. A 9 French sheath (Cordis, Johnson & Johnson, Florida, USA) was inserted, 20cc of contrast material was injected to obtain Inferior vena cavography and identify the level of stenosis.

A stiff hydrophilic guide wire (Radiofocus, Terumo, Japan) and a French vertebral catheter (Cordis, Johnson & Johnson, Florida, USA) were passed through the stenosis followed by dilatation with a non-complaint balloon (Atlas, Bard, USA) 18-20mm in diameter and 4cm in length inflated to up to 16 atmospheres. A completion venography was done to confirm patency of the IVC.

Manual compression of the puncture site was done.

In two cases, where the completion angiogram of the IVC post-venoplasty showed residual significant stenosis, a metal stent was indicated. The 9 French sheath was upgraded to 11 French. A self-expanding Wallstent (Boston Scientific, Ireland) 18 mm x 90 mm stent was placed. A completion venography was done to confirm patency of the IVC.

Manual compression of the puncture site was done.

II- Hepatic vein angioplasty and stent:

In eight cases, the IVC was patent, but there was stenosis at the level of the junction of the hepatic vein and IVC.

In these cases, an ultrasound guided transhepatic puncture of the patent hepatic vein was performed using 16 Gauge needle (Angiocath, Becton Dickinson, USA). A stiff hydrophilic guidewire is used, and a 6 French sheath is inserted. Contrast was injected to identify the stenosis. The guidewire is passed through the stricture. A 10mm x 4cm balloon (Mustang, Boston Scientific, Ireland) was used. A completion venography was done through the sheath with its tip within the hepatic vein to confirm a wide junction between the hepatic vein and the IVC.

In four cases, where the completion angiogram showed residual significant stenosis, a metal stent was indicated. A self-expanding Wallstent (Boston Scientific, Ireland) 10mm x 40mm stent was placed. A repeat completion angiogram was done.

In all eight cases, the sheath is removed, and a 5 French catheter is introduced over the guidewire and a 5mm 4cm metallic coil (Cook, USA) is introduced via the catheter with the liver tract superficial to the puncture site of the hepatic vein to avoid development of an intrahepatic hematoma.

III- Collateral hepatic vein angioplasty:

In one case, the IVC was patent, all three hepatic veins were occluded, but there was a large-caliber

intrahepatic collateral vein seen communicating with the IVC, showing a stenosis at the level of the junction with the IVC.

In this case, an ultrasound guided transhepatic puncture of the collateral hepatic vein was performed using Angiocath. A stiff hydrophilic guidewire is used, and a 6 French sheath is inserted. Contrast was injected to identify the stenosis. The guidewire is passed through the stricture. A 10mm x 4cm balloon was used. A completion venography was done through the sheath with its tip within the collateral hepatic vein to confirm a wide junction between the hepatic vein and the IVC.

The sheath is then removed, and a 5 French catheter is introduced over the guidewire and a metallic coil is introduced via the catheter with the liver tract superficial to the puncture site of the collateral hepatic vein.

Results

This is a retrospective study of Budd Chiari syndrome cases referred to our department for interventional radiology management in the period from September 2016 till August 2018. 25 cases of Budd Chiari syndrome were. We included the 14 cases of venoplasty who did not need a TIPSS procedure.

The current study included five males and nine females; their age ranged from 16 to 34 years with a mean age of 26.2 years.

In 13 cases, there was at least one non-totally occluded hepatic vein. One case of occluded hepatic veins, but patent intrahepatic collateral vein communicating with the IVC.

Distribution of cases according to Budd Chiari Classification:

- Type I: Membranous obstruction of the IVC W/WO thrombosis 5/14.
- Type II: Solitary hepatic vein obstruction (web/thrombosis) 8/14.
- Type III: Rudimentary (diffuse thrombosis) of hepatic veins 1/14.

Clinical presentation of the studied cases:

All patients presented with tense ascites; no patients presented with hematemesis. Four patients presented with dilated abdominal wall venous collaterals.

Techniques performed in the study:

- IVC balloon was done for three cases (Fig. 1).
- IVC balloon with stent was done for two cases (Fig. 2).
- Hepatic vein angioplasty was done for four cases (Fig. 3)
- Hepatic vein angioplasty with stent was done for four cases (Fig. 4).
- Collateral hepatic vein angioplasty was done for only one case (Fig. 5).

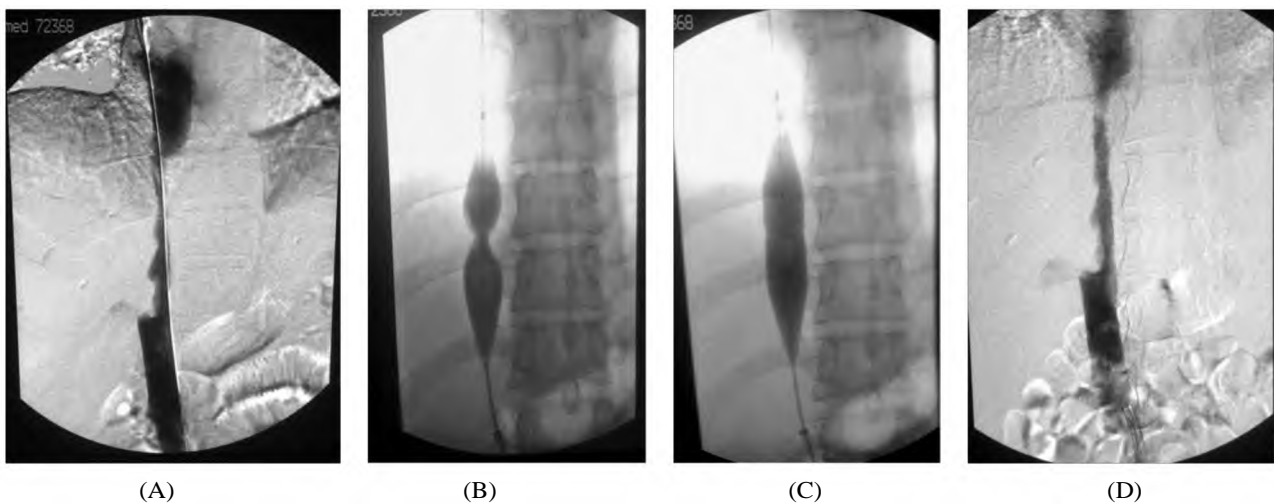


Fig. (1): IVC Balloon:

- A- Diagnostic venogram via femoral sheath showing stenosis of the hepatic IVC and partial thrombosis of the infrahepatic IVC.
 B- 18mm high pressure non-compliant balloon inflated showing a waist at the site of IVC stenosis.
 C- 18mm high pressure non-compliant balloon fully inflated.
 D- Completion venogram via femoral sheath showing free flow of contrast from the IVC to the right atrium with no residual focal stenosis.

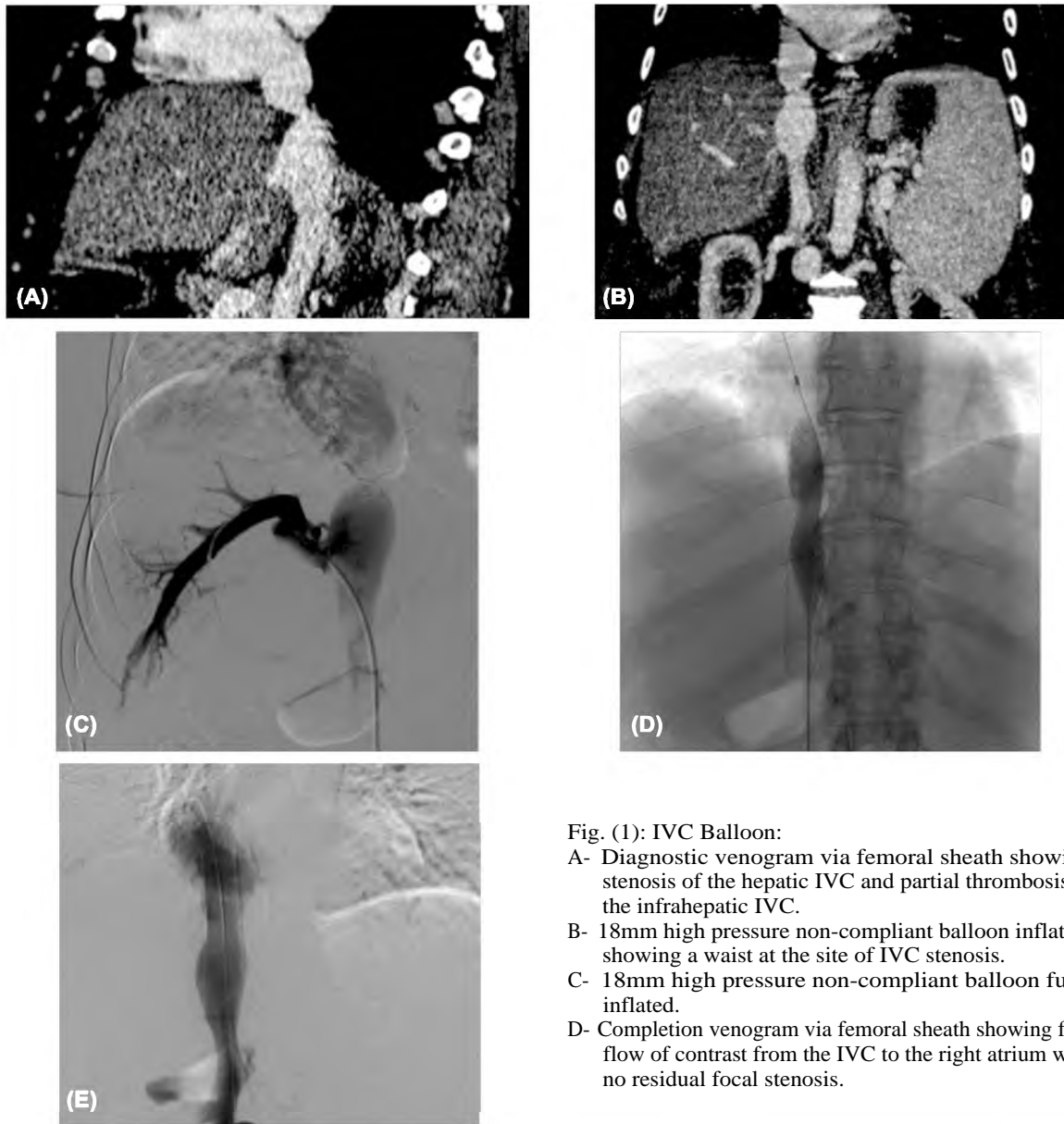


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- A- Diagnostic venogram via femoral sheath showing stenosis of the hepatic IVC and partial thrombosis of the infrahepatic IVC.
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- C- 18mm high pressure non-compliant balloon fully inflated.
- D- Completion venogram via femoral sheath showing free flow of contrast from the IVC to the right atrium with no residual focal stenosis.



Fig. (3): Hepatic vein angioplasty.

- A- Contrast is injected in the hepatic vein via an 16G needle through percutaneous transhepatic approach showing tight stenosis at the junction of the vein with the IVC with multiple small collaterals.
- B- 10 mm x 4 cm balloon angioplasty was done.
- C- Completion venogram revealed no residual stenosis.

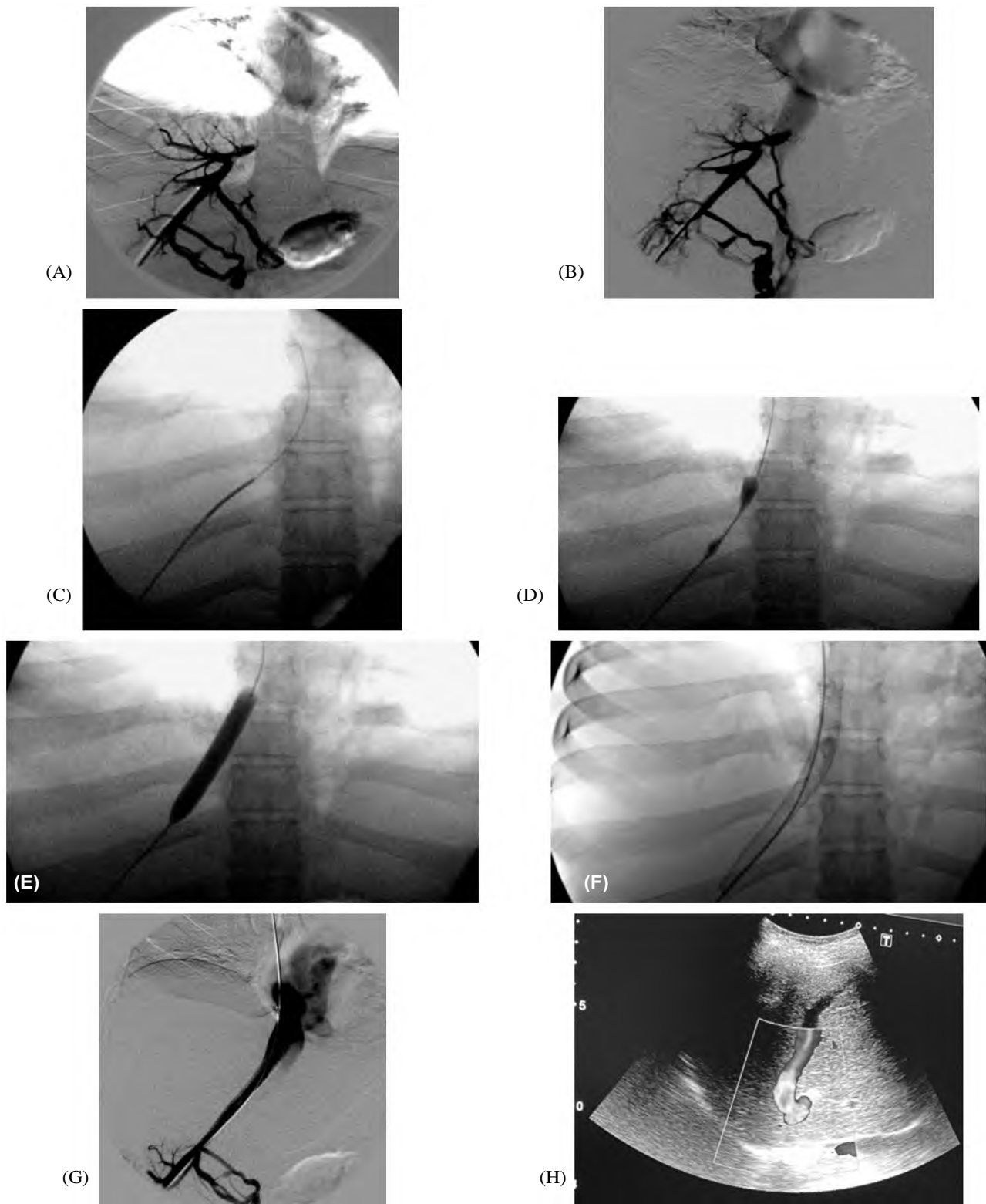


Fig. (4): Hepatic vein stent.

- A- Contrast is injected in the hepatic vein via an 16G needle through percutaneous transhepatic approach showing dilated hepatic vein with multiple large collaterals and no direct communication with the IVC.
- B- Delayed image of the previous venogram showing delayed opacification of the IVC with contrast mainly from the venous collaterals.
- C- A Cobra catheter and a guidewire is negotiated through the stenotic junction between the hepatic vein and the IVC.
- D- Partially inflated 10mm balloon showing a waist at the site of stenosis.
- E- Fully inflated balloon.
- F- A 10mm x 40mm stent is deployed.
- G- Completion venogram showing free flow of contrast through the stent with reduced collaterals.
- H- Follow-up Doppler examination after 3 years showing good flow within the hepatic vein and the stent.

Technical success rate was 100%, and after management of the cases all patients were put on life-time anticoagulant therapy and the INR is monitored between 2-2.5.

Follow-up ultrasound monitoring of the patency of the venoplasty and ascites was done after one week, one month and then every 6 months. As well as monitoring of liver function tests.

All liver function tests were improved including serum total bilirubin and serum Albumin. Albumin level mean increased from 2.5g/dl (2.1-2.9) to 3.3g/dl (2.8-3.5) after 7 months. Total bilirubin

level mean decreased from 2.4g/dl (1.9-2.8) to 1.7g/dl (1.4-2) after 7 months.

The follow-up period ranged from 18 to 72 months with no case of recurrent stenosis or occlusion was noted.

All patients had immediate improvement of ascites, 10 patients showed completely resolved ascites and 4 patients showed significant improvement with only minimal residual ascites.

The abdominal wall venous collaterals have disappeared in all patients.

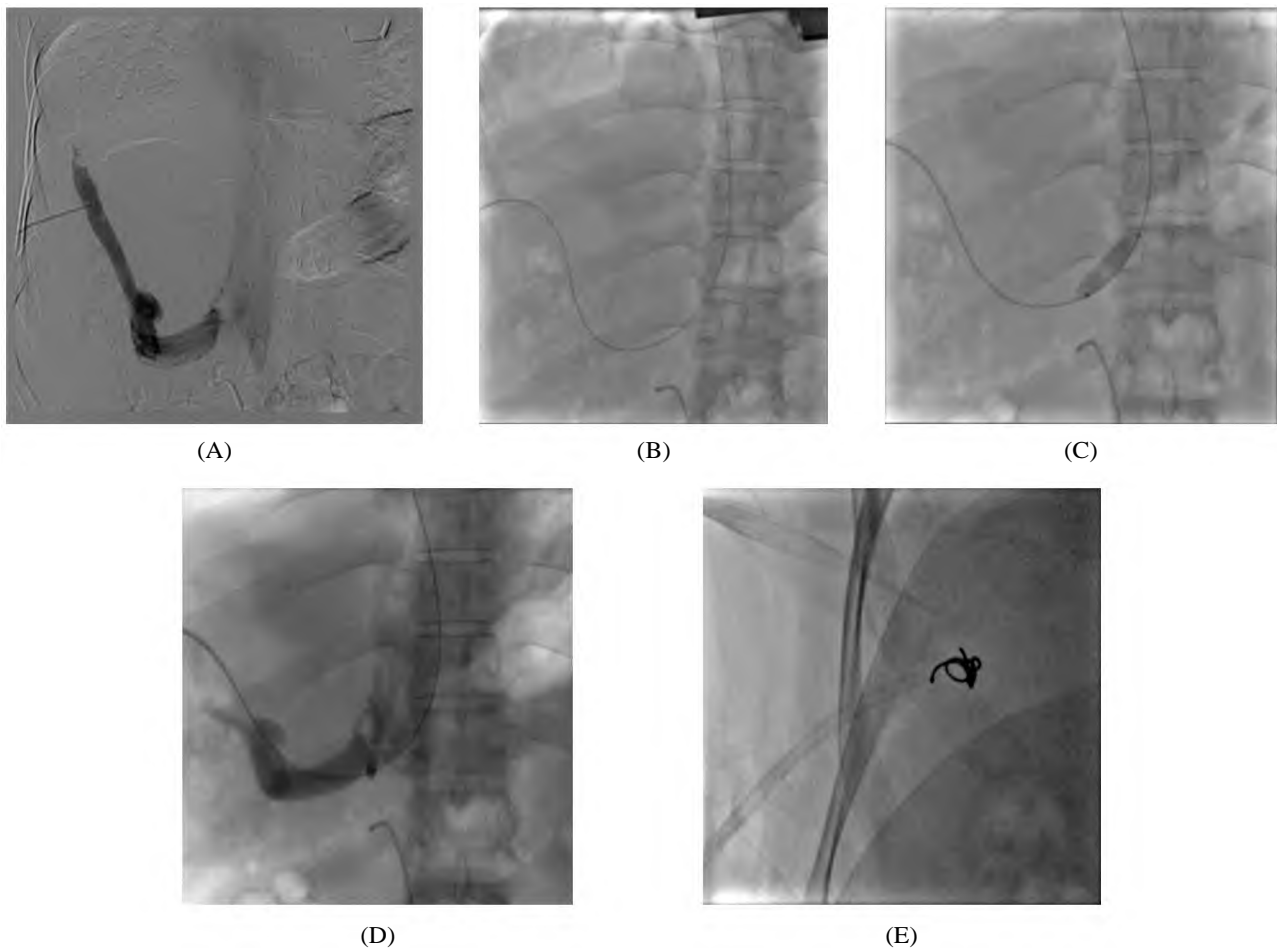


Fig. (5): Hepatic vein collateral angioplasty.

- A- Contrast is injected in the hepatic vein collateral via an 16G needle, through percutaneous transhepatic approach showing tight stenosis at the junction of the vein with the IVC with multiple small collaterals and faint opacification of the IVC.
- B- A guidewire is passed through the stenosis.
- C- 1.0mm balloon angioplasty was performed.
- D- Free flow of contrast from the collateral hepatic vein into the IVC with no residual stenosis.
- E- A metallic coil was deployed in the liver tract prior to withdrawal of the sheath to avoid bleeding.

Discussion

The first step in management of BCS is the use of anticoagulants with medical treatments, endoscopic and/or medical management of the related hepatic complications, recanalization of the stenotic vein(s) using venoplasty with or without stenting according to the condition to restore normal hepatic flow and relieve the congestion, if these techniques failed, TIPSS and then liver transplantation are the treatment of choice [8].

The main target in treatment of BCS is to relieve hepatic congestion in order to avoid poor outcome with progression of hepatic decompensation, failure and death [2].

Radiological intervention nowadays plays a major role in management of BCS in the form of recanalization of occluded veins using different techniques as balloon dilatation with or without stenting and TIPSS [2,5,7].

After different studies, that showed the efficacy of interventional radiology in management of BCS patients, interventional radiology nowadays is preferred than surgical shunts with better prognosis [2,9].

Our aim was to assess the efficacy of venoplasty in management of patients with BCS and actually we detected high success rate among the managed cases with satisfactory follow-up to more than 6 years in some cases with no evidence of recurrence, this was in agreement with Yonghua Bi, et al., [7] as they also confirmed high success rate among their cases after venoplasty. Nitin Jagtap, et al., [10] confirming the added value of angioplasty as a treatment of BCS cases with higher success rate than previous studies. Also, Qiuhe Wang, et al., [11] concluded that angioplasty with stenting is more superior than angioplasty alone, mainly in cases with short length stenosis, with better outcome.

The role of Doppler ultrasound in diagnosis of BCS regarding the occluded hepatic veins with/without IVC involvement is of great help in diagnosing the type, extent and cause of obstruction with high sensitivity and accuracy, not only that but it helps to depict the presence of collaterals which can change the management of the case. Varun Bansal, et al., [5] and Simón Correa Gaviria, et al., [12] mentioned that Doppler US plays an important role in diagnosis of BCS and considered the first imaging modality of choice in these cases also to detect the associated collaterals with high sensitivity and specificity up to 85%. Doppler

examination was an integral part of patients' assessment and management plan in our cases.

Thomas Mammen, et al., [13] concluded that it is important to identify the presence of intrahepatic collaterals in BCS cases, which helps in management of these cases by recanalization of these occluded/stenotic collaterals and considering it a promising technique in management of cases with failed hepatic veins recanalization and recommend further studies on large number of cases. This was in agreement with our study, although we had only one case of stenotic intrahepatic collateral with occluded three hepatic veins and a patent IVC; angioplasty was done for dilatation of the stenotic collateral with good outcome on long-term follow-up for more than 6 years. We recommend the use of this technique on a larger number of cases in future studies; however, this needs thorough Doppler examination of BCS patients to diagnose the presence of such collaterals in direct communication with the IVC. As to our knowledge, there are no enough studies that discuss this point.

Yonghua Bi, et al., [14] recommended the use of TIPSS in cases of failure of angioplasty, actually in our cases we did not face such condition as we had 100% technical success rate.

Conclusion:

Budd Chiari patients should have a thorough Doppler examination to assess the presence of a communication between the hepatic vein or their intrahepatic collaterals with the IVC. The presence of this venous communication could allow us to perform a venoplasty with/without stenting and avoid performing unnecessary TIPSS procedure. Venoplasty has a very good long-term outcome in Budd Chiari syndrome.

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علاج متلازمة باض كيارى دون التحويلة البابية الجهازية داخل الكبد عبر الوداجى

متلازمة (BCS) هي مجموعة غير متجانسة من الاضطرابات الكبدية الوعائية. يلعب التصوير مع التطورات الحديثة في هذه التقنيات دوراً مهماً في تشخيص وإدارة الحالات مع BCS. كان الهدف من العمل الحالي هو دراسة الفعالية السريرية لعملية رأب الوريد، دون الحاجة إلى إجراء TIPSS، في إدارة BCS.

النتائج: سريرياً، تم تضمين جميع مرضى BCS الذين ظهرت عليهم أعراض آلام في البطن وانتفاخ و علامات تضخم الكبد والاستسقاء. تم أخذ الموافقة المسبقة من جميع المرضى. تمت معالجة هذه الحالات على النحو التالي: ثلاث حالات: رأب الوريد الوريدي حالتان: رأب الوعاء الدموي والدعامات. أربع حالات: رأب الوريد الكبدى. أربع حالات: رأب الوريد الكبدى والدعامات حالة واحدة: رأب الوريد الكبدى الجانبي

الخلاصة: يجب أن يخضع مرضى BCS لفحص دوبلر شامل لتقييم وجود اتصال بين الوريد الكبدى أو الضمانات دخل الكبد مع IVC. يمكن أن يسمح لنا وجود هذا الاتصال الوريدي بإجراء عملية رأب الوريد مع / بدون دعامة وتجنب إجراء TIPSS غير الضروري. تحقق عملية توسيع الوريد نتائج جيدة جداً على المدى الطويل في متلازمة BCS.