

Original Article

Early experience of Budd-Chiari syndrome treatment with transjugular intrahepatic portosystemic shunt

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Abstract

The Budd-Chiari syndrome is a heterogeneous group of disorders characterized by obstruction of hepatic venous outflow at any level from the small hepatic veins to the junction of the inferior vena cava with the right atrium. We present two cases of Budd-Chiari syndrome with severe ascites associated with polycythemia vera in first case and protein C deficiency in the second, in both cases transjugular intrahepatic portosystemic shunt were placed, with excellent control of symptoms, no mortality were observed, and just one episode of pulmonary venous thrombosis was observed. To our knowledge this is the first time that transjugular intrahepatic portosystemic shunt are used and reported in Budd-Chiari syndrome in Mexico.

Key words: Budd-Chiari syndrome, TIPS, complications.

BACKGROUND

Budd-Chiari syndrome (BCS) is an uncommon liver disease defined as an obstruction to hepatic venous outflow at any level from the small hepatic vein to the junction of the inferior vena cava and the right atrium. Heart failure and sinusoidal obstruction syndrome (formerly

known as veno-occlusive disease) also impair hepatic venous outflow and share many features with BCS, but are considered separately as causes and treatments are different.¹ The obstruction of the hepatic venous outflow leads to sinusoidal congestion, centrilobular necrosis, fibrosis and portal hypertension² as consequences variceal bleeding, refractory ascites, hepatorenal syndrome and hepatopulmonary syndrome are very severe complications of BCS, with a potentially fatal outcome if no treated appropriately.³⁻⁵

Although the natural history of BCS is not well known, mortality is highest at first 2 years of diagnosis and was found to be independent of surgical portosystemic shunting⁶ and is affected by several prognostic indicators as the severity of encephalopathy, ascites, prothrombin time and serum levels of bilirubin.⁵

The main standard of treatment is based in anticoagulation unless contraindicated, but anticoagulation soled treatment is associated with high mortality at 6 months,⁷ considering this information thrombolytic therapy or shunting is indicated according to an individualized care.

Transjugular intrahepatic portosystemic shunt (TIPS) have been shown to be an efficient portal-systemic derivative treatment for BCS patients uncontrolled by medical therapy. TIPS overcomes sinusoidal congestion, caudate lobe and inferior vena cava compression, with less proceeding-mortality associated than surgical shunting, particularly in patients with poor liver function and inferior vena cava compression.⁸ Besides, after successful decompression stent dysfunction is not necessary associated with worsening portal hypertension⁹ which could be explain by intrahepatic collateral circulation development so when TIPS dysfunction occur the hepatic outflow could be made by collateral circulation. However, the main drawback of TIPS for this condition is a very high rate of shunt dysfunction with uncovers stents. Recently, polytetrafluoroethylene-covered stents have been shown to reduce the incidence of TIPS dysfunction in BCS patients.¹⁰

The aim of this paper is describe our experience in treatment of BCS with TIPS, a technique not previously made in Mexican population with BCS.

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Case presentation

Case 1

A forty-eight years old female, with previous lose of pregnancy in the first trimester and superficial venous thrombosis

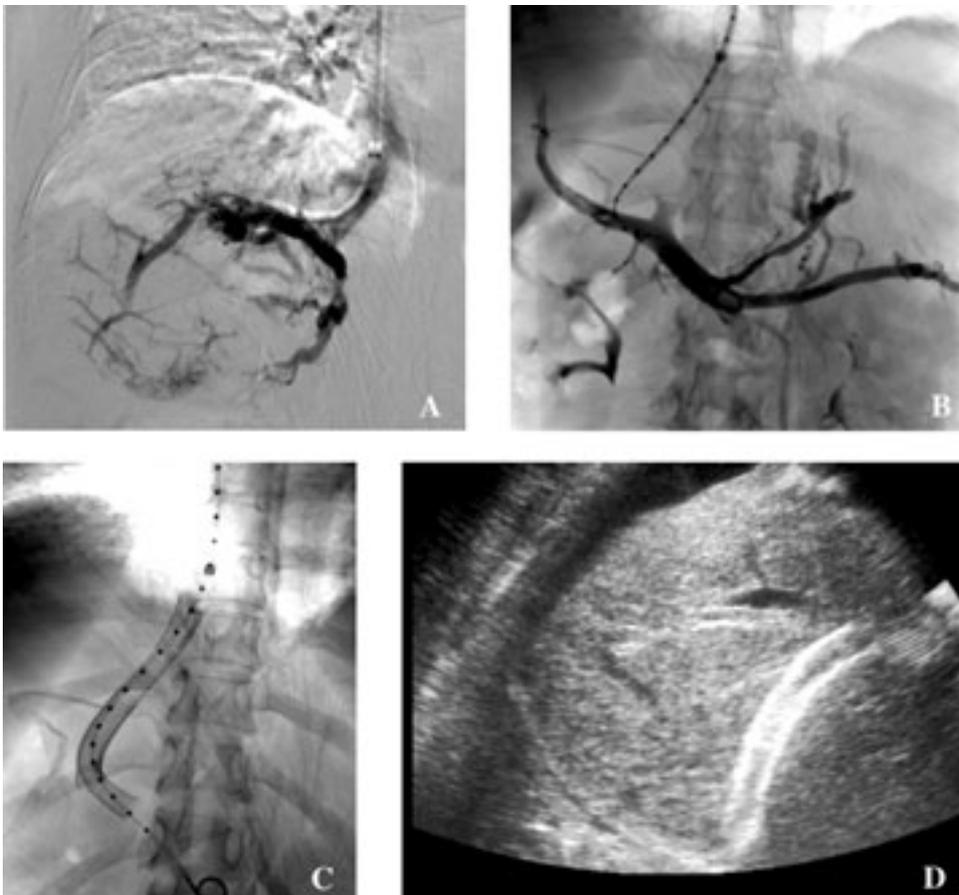
Table I. Characteristics of patients with Budd-Chiari syndrome.

	Patient 1	Patient 2
Age	48	17
Gender	Female	Male
Underlying disease	Polycythemia vera	Protein C deficiency
Clinical presentation		
Ascites	+	+
Hepatic encephalopathy	-	-
Upper gastrointestinal bleeding	-	-
Clichy score	6.1	5.1
Rotterdam score	1.04	1.04
Complications	Pulmonary thromboembolism	None
Mortality	None	None
Porto Caval pressure gradient pre-TIPS (mmHg)	35	42
Porto Caval pressure gradient post-TIPS (mmHg)	15	20
% Change in gradient	- 43%	- 48%

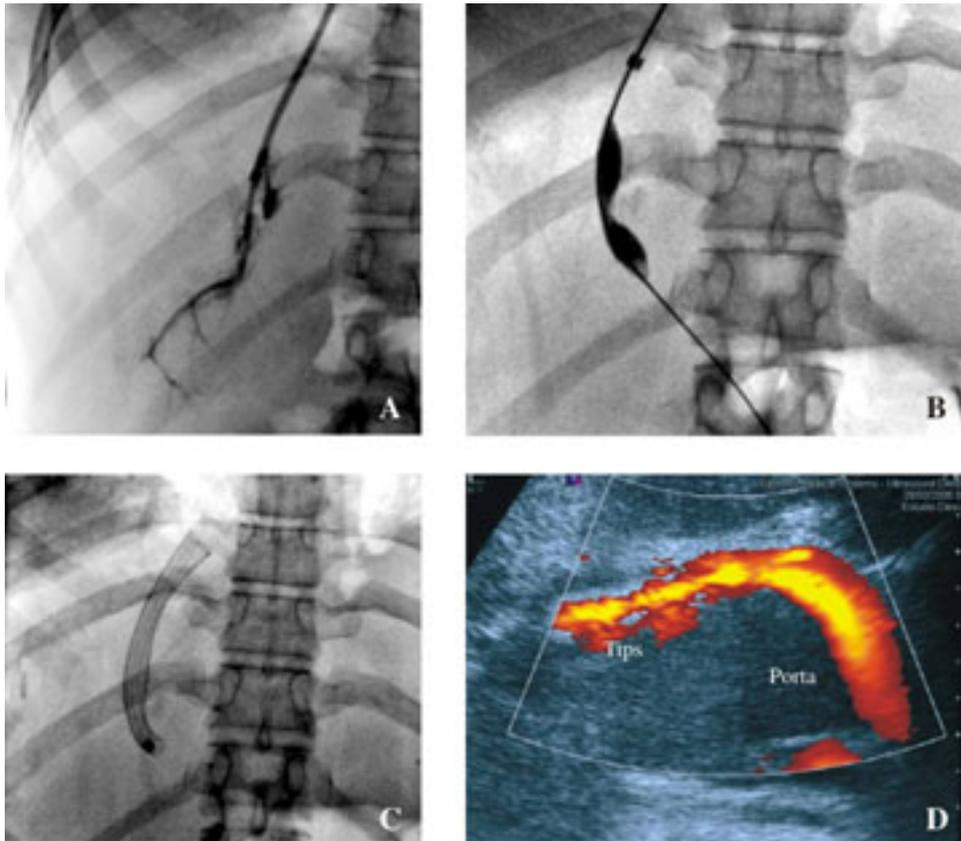
of lower left limb. Her symptoms started three months before she gets to our hospital, she referred progressive ascites and edema on lower limbs with abdominal collateral circulation. Large esophageal varices were detected at endoscopy. Ultrasonographic Doppler images and computed tomography suggest suprahepatic vein and retrohepatic inferior vena cava thrombosis. The venography confirmed thrombosis of the three hepatic veins (Case 1 Images). Polycythemia Vera was diagnosed. Because a refractory ascites was developed a non-covered TIPS (2 wall stents, Boston Scientific) were placed through the proximal portion of the right hepatic vein. Portocaval pressure gradient was reduced from 35 mmHg to 15 mmHg (Table I). However, after TIPS placement the patient developed dyspnea, a high-definition chest tomography diagnosed right side subsegmentary pulmonary thromboembolism and a subcapsular liver hematoma was detected. Greenfield filter was placement, after three days of careful observation with non-invasive mechanical ventilation and anticoagulation, the patient was discharged home without other complications. During follow up the ascites has disappeared and the patient does not require diuretic therapy.

Case 2

A seventeen years old male, with family history of thalassemia in a sister, he was diagnosed previously as



Images Case 1. A) Thrombosis of the hepatic vein, collaterals around a nodule. B) Puncture from the right hepatic vein stump to the right portal branch vein. C) TIPS (2 wall stent, Boston Scientific). D) TIPS image by ultrasound.



Images Case 2. **A)** Abnormal flow of the hepatic vein. **B)** Balloon dilatation of the liver parenchyma tract. **C)** TIPS (wall stent, Boston Scientific). **D)** TIPS image by Doppler color ultrasound.

thalassemic trait. Three months before a complain of pain in upper right quadrant and early satiety progress to important increase of abdominal circumference and vascular spiders. Physical examination demonstrates jaundice, severe ascites and hepatomegaly, large esophageal varices were detected. Doppler ultrasound and computed tomography can not exclude portal or hepatic vein thrombosis. A diagnostic venography was performed and showed suprahepatic vein thrombosis of the three veins, in second time an uncover-TIPS (Wall-Stent, Boston Scientific) was inserted through the proximal portion of the right hepatic vein (Case 2 Images), portocaval pressure gradient was reduced from 42 mmHg to 20 mmHg, no complications were observed after TIPS placement and resolution of ascites was observed after 48 hours (Table I), investigation about thrombophilia demonstrated protein C deficiency so anticoagulation therapy was initiated.

Discussion

At the end of 90's decade the standard treatment options for patients who presented with BCS were medical management, surgical shunts, or liver transplantation, unfortunately medical management with diuretics, anticoagulants, and paracentesis has been shown to be ineffective with respect to survival.^{7,11} Surgical shunts are performed mainly to relieve the obstruction causing portal hyperten-

sion, they derivate the portal flow to others territories as inferior cava vein in the case of portocaval shunt, unfortunately the success of these shunts depends of portocaval gradient and as others authors have described a portocaval pressure gradient > 10 mmHg is required to success portocaval shunt because of frequent inferior vena cava compression in BCS.¹² Liver transplantation is indicated for liver failure.

In this issue we report the early experience in treatment of BCS with TIPS. To our knowledge this is the first communication about this therapeutic modality in our country, especially if we consider that BCS is a rare disease even in specialized centers.¹³ In this initial experience we observe excellent outcomes, with similar results to other more experienced groups,¹⁴ all TIPS were successfully placed and no mortality or major morbidity associated.

Although, because our short follow up (five months) we still can not evaluated survival impact. The expected actuarial survival at 1 year and 5 years reported in BCS and TIPS treatment is of 94 % (95% CI 88 – 100%) and 87 % (95% CI 78 – 96%) respectively,^{9,13} indicating that use of this strategy is safe and useful in experienced groups.

After TIPS placement an excellent reduction in portal pressure was observed in this and others series.¹⁵ The successful outcomes in our series are influenced by good general status, one of the most important prognostic

factor.¹³ At five months of follow up there is no evidence of TIPS dysfunction in both patients evaluated by Doppler ultrasound (performed monthly in the first three months) and both of them are clinically asymptomatic with excellent ascites remission. We used uncovered stents for economical reasons as we know the very high rate of dysfunction with uncover stents that can be reduce with covered stents with a final economic impact throw the follow up. TIPS placement in a hepatic thrombosis vein is technically possible from a puncture of the right hepatic vein stump to the right portal branch vein, as the present two cases, another approach could be a direct puncture from the intrahepatic portion of the inferior vena cava to the right portal vein. There is no data about the optimal reduction of portocaval pressure gradient after TIPS placement in BCS patients, as the optimal portocaval pressure gradient that needs to be obtained for the control of refractory ascites associated with cirrhosis is even less clear, different for variceal bleeding, (cut off 12 mmHg), some authors suggested less than 8 mmHg for refractory ascites.^{16,17} In the present both cases we obtained more than 40% of reduction of the portocaval pressure gradient, none get < 12 mmHg but there was excellent control of ascites.

In conclusion this report shows that in our population TIPS treatment in BCS patients is safe and successfully.

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