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Living donor liver transplantation for adult Budd Chiari syndrome – Resection without replacement of retrohepatic IVC: A case report

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ABSTRACT

INTRODUCTION: Suprahepatic caval resection and replacement of inferior vena cava (IVC) is standard procedure in deceased donor liver transplantation for patients with Budd-Chiari syndrome (BCS). However, replacement of IVC in living donor liver transplantation (LDLT) is difficult. We report a case of BCS successfully treated by LDLT without replacement of IVC.

PRESENTATION OF CASE: A 52-years-old female with a primary BCS due to IVC thrombosis. A vena cava (VC) stent placed after angioplasty without improvement of the hepatic, portal venous flow and liver functions, Transjugular intrahepatic portosystemic shunt was considered and the patient had a rapid deterioration and increased ascites. The patient was scheduled for living donor liver transplantation (LDLT). Her Child-Paugh and MELD scores were 11, 18, respectively at time of transplantation. Left lobe was obtained from her son. Preservation of the native suprarenal IVC was impossible due to massive fibrosis and thrombosed. The suprahepatic IVC was also fibrotic and unsuitable for anastomosis with hepatic vein. The retrohepatic IVC resected include suprahepatic IVC together with the liver. The supradiaphragmatic IVC was reached and encircled through opening the diaphragm around the IVC and a vascular clamp applied on the right atrium with subsequent anastomosis with hepatic vein of the graft. The hemodynamic stability of the patient was maintained throughout the operation without IVC replacement due to developed collateral vessels.

CONCLUSION: Patients with Budd-Chiari syndrome with obstructive IVC are successfully treated with living donor liver transplantation without replacement of IVC.

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1. Introduction

Despite being a definitive treatment of Budd-Chiari syndrome (BCS), liver transplantation (LT) indicated when patients with BCS had liver failure, advanced cirrhosis or had a failed attempt of portosystemic shunt and other non-surgical therapeutic modalities [1]. In deceased LT, since the graft contains the retrohepatic IVC, the recipient IVC replaced by that of the donor. In contrast to living donor Liver Transplantation (LDLT), where the venous reconstruction is more difficult and challenging. Innovations in sur-

gical techniques and venous models were generated to overcome these difficulties either by preserving the native IVC or replacing it with homologous or synthetic vein grafts [2,3]. However, etiological point of view, one of the causes of Budd-Chiari syndrome is morphological abnormality including stenosis of suprahepatic IVC that should be removed to prevent recurrence of disease.

In our patient we could not preserve the recipient retrohepatic IVC including suprahepatic IVC due to fibrosis and thrombosed. Herein we described innovative technique for hepatic venous reconstruction for a patient with Budd-Chiari syndrome who needs to remove suprahepatic IVC in LDLT. This article has been written in line with the SCARE criteria as described by Agha et al. for the SCARE group. "the SCARE statement: consensus-based surgical case report guidelines. International journal of surgery 2016 [4].

Abbreviations: BCS, Budd-Chiari syndrome; CT, computed tomography; HV, hepatic vein; DUS, Doppler ultrasonography; IVC, inferior vena cava; LT, liver transplantation; LDLT, living donor liver transplantation; PV, portal vein; TIPS, transjugular portosystemic shunt.

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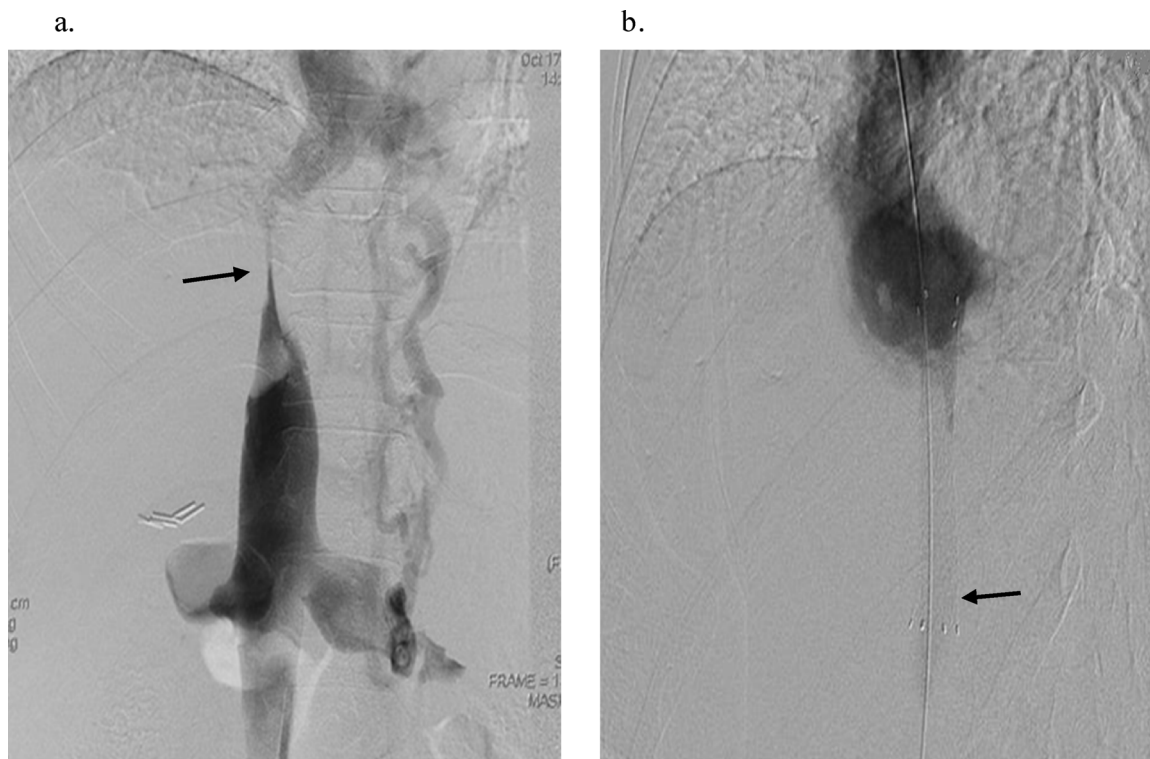


Fig. 1. inferior vena cography reveals thrombotic portion of the IVC (arrow) before (a) and after (b) metallic stent placement (arrow).

2. Case presentation

A 52-year-old female who had no past medical history and no family history had systemic edema and ascites. Computed tomog-

raphy (CT) and Doppler ultrasonography (DUS) revealed thrombus and obstructing the IVC and hepatic veins. Budd-Chiari syndrome was diagnosed with these findings. Percutaneous transluminal

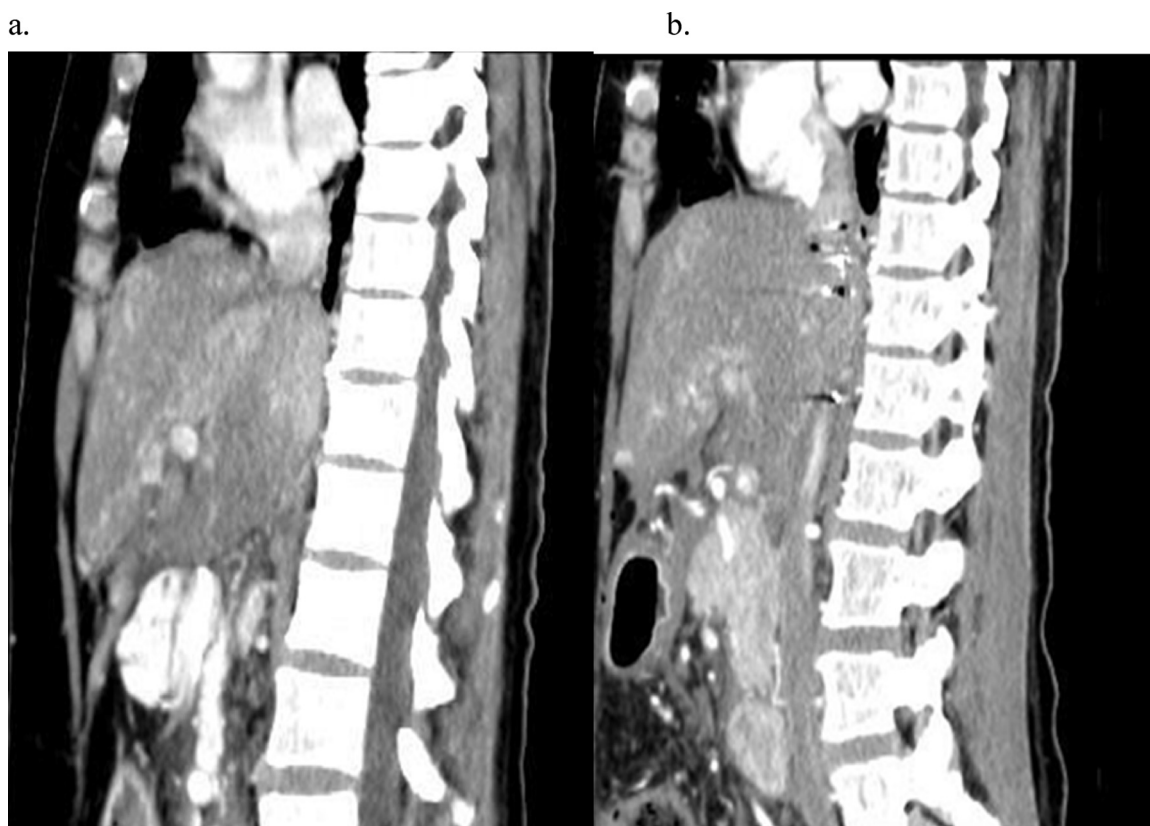


Fig. 2. Sagittal Computed tomography (CT) showing absence of flow in the hepatic veins before (a) and after (b) IVC stent placement.

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