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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-

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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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 SCARCE group. “the SCARE statement: consensus-based surgical case report guidelines. International journal of surgery 2016 [4].

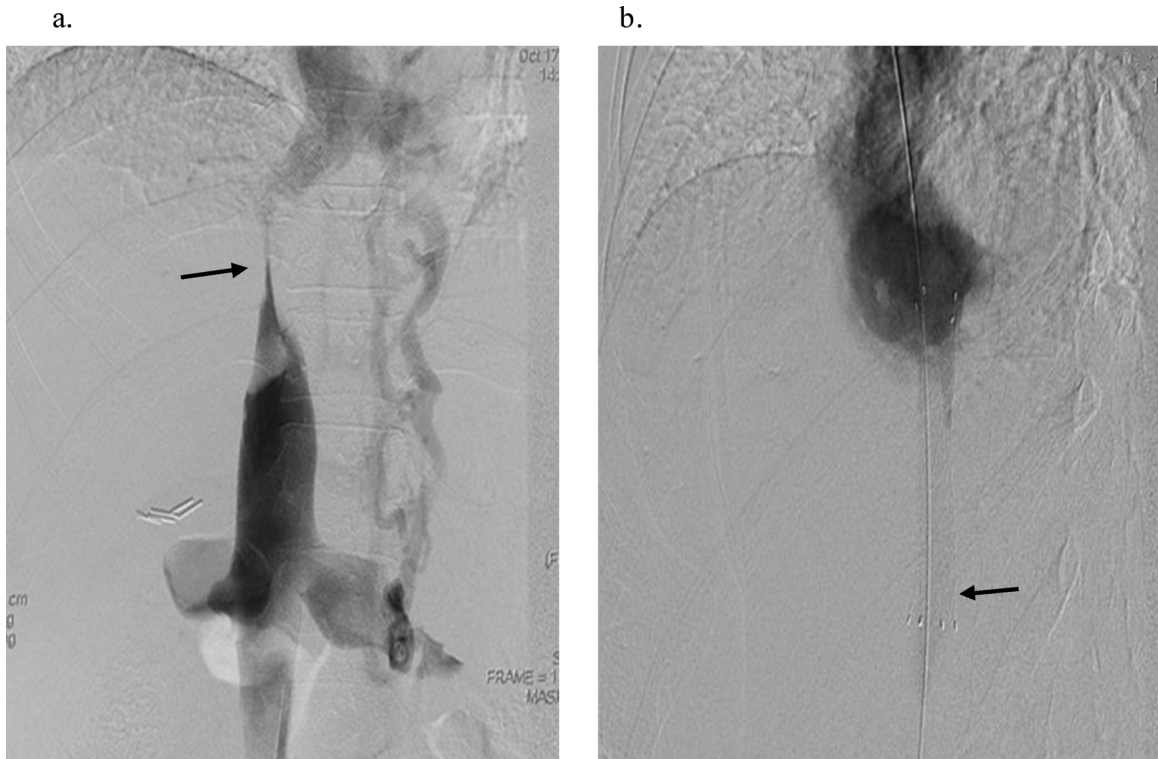


Fig. 1. inferior vena cography reveals thrombotic portion of the IVC (arrow) before (a) and after (b) metallic stent placement (arrow).

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.

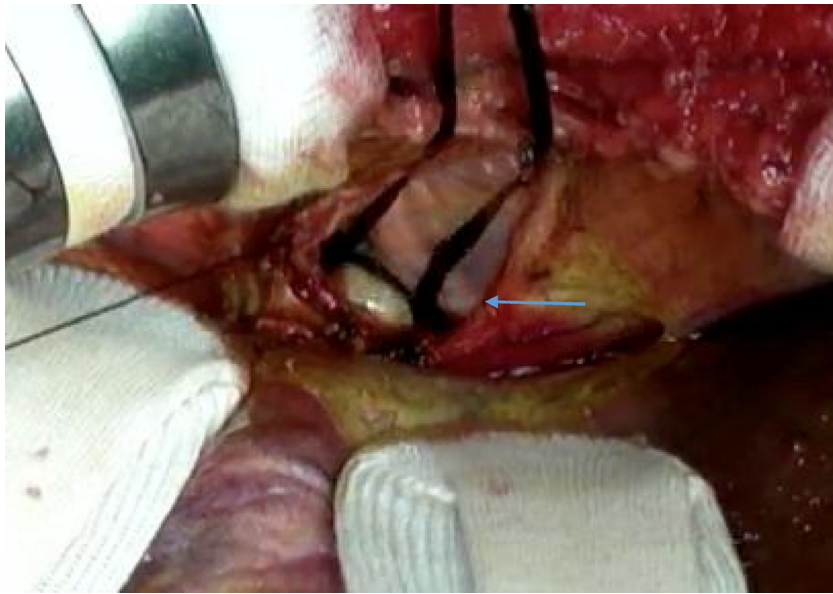


Fig. 3. Encircling the supradiaphragmatic IVC by vessel loop through a window made in the diaphragm (blue arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

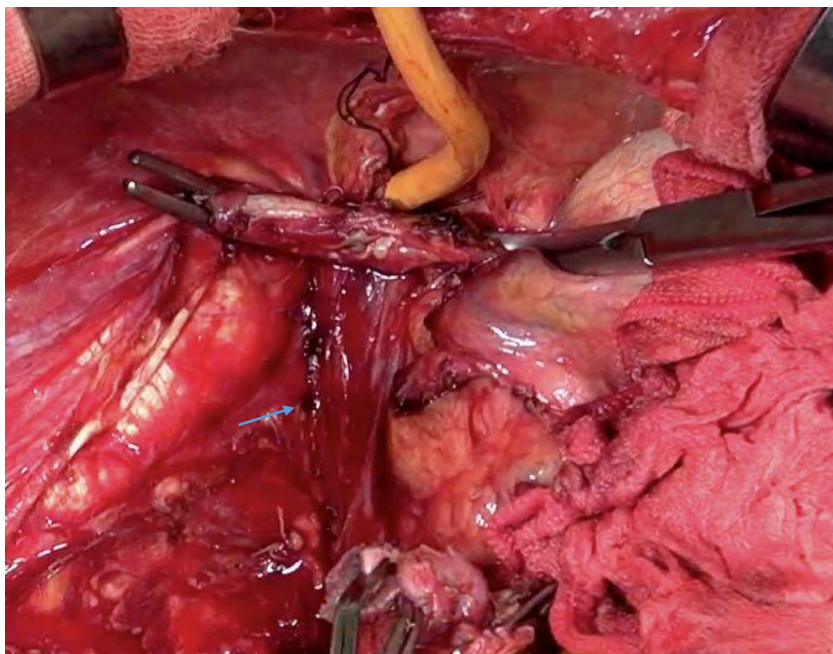


Fig. 4. Total hepatectomy together with excision of the retrohepatic IVC (blue arrow). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

angioplasty was performed with thrombectomy and IVC stent placement by the intervention radiology team and anticoagulants treatment for hepatic vein obstruction was performed as the initial management (Fig. 1). Despite these treatments, portal and hepatic venous flow by UDS, symptoms such as systemic edema and ascites and liver functional tests were not improved. Transjugular intrahepatic portosystemic shunt (TIPS) was considered as a second therapeutic option. However, it had failed and had increased ascites with rapid deterioration of liver functional tests such as the serum levels of bilirubin 8 from 4 mg/ml and INR 2.2 from 1.2 within one month. Liver transplantation was considered and the patient was referred to our hospital. The patient was scheduled for liver transplantation. The Child-Paugh and the MELD scores were 11,

18, respectively at the time of transplant qualification. Preoperative enhanced abdominal CT scan obtained stenosis of distal end of IVC stent, abundant ascites, a thrombus in the transverse portion of the left portal vein, and could not identify any of the hepatic veins.

To determine the cause of BCS; protein C and S activities, and Gene mutations for thrombosis were assessed and did not assign specific cause. However, Bone marrow biopsy findings were consistent with polycythemia vera with Hb level 10.6 gm/dl.

A left lobe graft liver with 512 gm weigh, a single orifice left; hepatic artery, portal vein, bile duct, was obtained from patient's 21-year-old son, the body mass indices of the recipient and donor were, 22.5, 24.5 respectively, with GWRW% 0.8. There were no abnormalities in the pretransplant work up with identical ABO

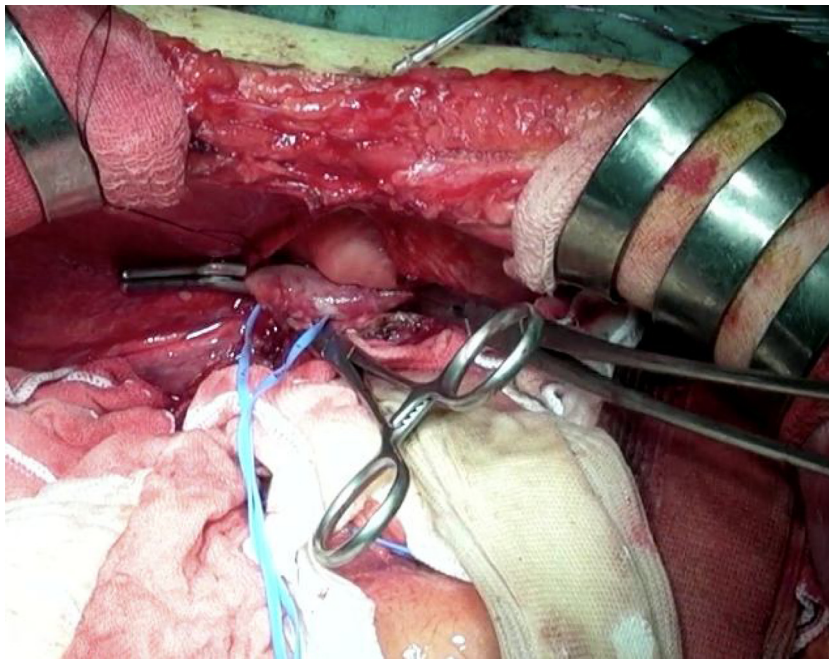


Fig. 5. Clamp applied on the RT atrium bottom to prepare it for the hepatic venous outflow reconstruction.

blood type histocompatibility. The period between the onset of the syndrome and LT was 2 months.

Liver transplantation was performed with taking informed consent from donor, recipient and families. Surgery for the donor and the recipient had been stated elsewhere [5]. In BCS cases, diffuse fibrosis of the retroperitoneum involving the IVC and the previously inserted a stent make mobilization difficult [6,7]. We encircled the supradiaphragmatic IVC after opening the diaphragm around the vena cava longitudinally with diathermy for hepatic vein anastomosis (Fig. 2). Cross clamps were applied at supradiaphragmatic IVC and suprarenal IVC. The recipient cirrhotic liver was totally explanted with the retrohepatic IVC (Fig. 3). Then, we used the right atrium for the outflow reconstruction, the vascular clamp was applied on the right atrium through the window made in the diaphragm (Fig. 4). After the anesthesiologist ensured absence of cardiac arrhythmia and hemodynamic abnormality, the atrium was cut about 2.5 cm at the bottom to match the graft HV, 5/0 polypropylene suture used for the anastomosis between the atrium and HV of the graft liver in a continuous fashion. A 24 Fr drain tube was inserted at the right pleural space. The window in the diaphragm around the HV-atrium anastomosis was minimized without complete closure, to avoid collection of cardiac effusion. Further steps of transplantation were performed in a standard procedure, except for the PV where vein graft interposition obtained PV from the explanted liver for reconstruction. The graft liver placed more cranial side than standard liver transplantation because HV anastomosis was performed above the diaphragm. So that for PV reconstruction, vein graft interposition was necessary due to longer distance between PV of the graft liver and PV of the recipient. Hemodynamic stability was maintained throughout the operation without veno-venous bypass that means enough collateral vessels were developed in systemic venous return. Operation time was 13 h 51 min and blood loss was 12150 ml.

The patient spent 8 days in the intensive care unit with uneventful course. Postoperative DUS confirmed hepatic venous patency with triphasic flow, and echocardiogram showed no abnormalities. Immediate postoperative anticoagulant therapy in form of IV heparin was adjusted to maintain INR at 2. The standard immunosuppressive regimen with tacrolimus was used. The patient was

recovered from surgery on the POD 35 and discharged from the hospital on the POD 80 after rehabilitation with oral anticoagulant warfarin concomitantly with acetylsalicylic acid in addition to hydroxycarbamide. Computed tomography 3 months after transplant confirmed anastomosis patency (Fig. 5). The patient has been doing well with normalized liver functional tests without post-transplant complications and recurrent disease such as rejection and other vascular complications.

3. Discussion

Budd-Chiari syndrome is a rare life threatening disorder, in which obstruction of the hepatic venous flow occurs at any level from the intrahepatic small veins to the right atrium, with exclusion of right heart failure and constrictive pericarditis [8]. It is a multifactorial disorder, with etiological factors differ among eastern and western countries. Unlike Asian countries where membranous obstruction of the suprahepatic IVC predominate [9], polycythemia vera was the associated etiology in our patient. Which was diagnosed by bone marrow biopsy findings, despite normal peripheral blood cell counts.

Symptoms and physical signs are not specific for BCS. However, it should be suspected in patients with tender hepatomegaly and refractory ascites without liver failure. Diagnostic imaging plays the main role in the diagnosis of BCS, DUS is considered the modality of choice in suspected cases with a high sensitivity and specificity, it can determine the site of obstruction and venous flow pattern [10]. Computed tomography and magnetic resonant imaging confirm the diagnosis and can exclude other causes with better visualization of liver parenchyma. Venography is considered the reference for the evaluation of the IVC and HVs, however, its invasiveness restricted its use for venous pressure measurement [11,12].

Liver transplantation should be a rescue modality after failed medical treatment such as anticoagulant therapy and TIPS [13]. However, the correct timing and the efficacy of each therapy are still controversial. In our case, we adopted the step by step therapy [13], and LT considered when there was no response to the other treatment strategies with rapid deterioration of liver func-

tional tests. The Child and MELD scores were 11 and 18, respectively at the time of transplantation.

As, the native IVC replacement is feasible in deceased donor liver transplantation. However, due to scarcity of the deceased donor programs in Asian countries, LDLT has become the main transplant procedure for patients with BCS. Different techniques were reported to overcome the technical challenges of the venous outflow reconstruction in LDLT for BCS. These include; patch plasty of the native IVC, replacement or interposition of the IVC using vein grafts, and direct anastomosis of the hepatic vein to rarely preserved IVC, suprahepatic IVC or IVC interposition [2,14]. Herein, we resected the suprahepatic IVC and anastomosed to right atrium, without replacement IVC because we considered more curative procedure to avoid recurrent disease due to membranous obstruction of the suprahepatic IVC. In fact, the suprahepatic IVC was fibrotic and stenotic in our case. The HV anastomosis to the right atrium allowed venous outflow without membranous obstruction of the suprahepatic IVC that was already removed.

The post-transplant outcome was uneventful without reconstruction of IVC due to well-developed collateral vessels that allowed enough systemic venous return. Removal retrohepatic IVC with suprahepatic IVC and without IVC replacement is safety procedure and successfully performed for BCS patients with obstructive IVC in living donor liver transplantation.

4. Conclusion

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

Conflicts of interest

No conflict of interest.

Sources of funding

We have no supportive funding.

Ethical approval

This work was approved by the Kyoto University ethics committee.

Consent

We obtained patient consent for publishing her case

Author contribution

Tarek Abdelazeem Sabra is a major contributor in writing the manuscript. Shinji Uemoto, Hidaeki Okajima, Kentaro Yasuchika, Koichiro Hata, Ken Fukumitsu, Tetsuya Tajima, Tarek Abdelazeem Sabra performed clinical treatment including surgical operation. All authors have read and approved the final manuscript.

Registration of research studies

This work does not require a registration of research studies because it is a case report.

Guarantor

Hideaki Okajima
Tarek Abdelazeem Sabra

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