

Liver and Partial Atrium Transplantation for Chronic Budd-Chiari Syndrome

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A 22-year-old man was admitted to our hospital for progressive liver failure with ascites, jaundice, and bleeding from esophagogastric varices due to chronic Budd-Chiari syndrome and antiphospholipid syndrome.

Nine years before, a transjugular intrahepatic portosystemic shunt (TIPS) had been placed between the right hepatic vein and the portal vein at another hospital. The patient subsequently developed acute obstruction of the hepatic veins followed by the progressive upward extension of the thrombus, requiring positioning of a second TIPS in the vena cava. Because of complete occlusion of both shunts, the patient developed the symptoms of chronic Budd-Chiari syndrome despite collateral circulation.¹⁻³

At our hospital, a computed tomography scan confirmed the presence of a thrombus in the vena cava from the renal veins to the right atrium (Fig. 1). The patient was scheduled for liver and partial right atrium transplantation with cardiopulmonary bypass.

The abdominal incision started when the ascending aorta, the superior vena cava, and the right femoral vein had been cannulated and normothermic cardiopulmonary bypass had been instituted. This strategy helped us to decompress the portal vein and the collateral circulation and permitted an easier approach to the right atrium. The recipient hepatectomy was performed with the resection of the vena cava from the renal veins to the right atrium and with the opening of the diaphragm through the central fibrous body

(Fig. 2). Occlusion of the caval lumen up to the right atrium was confirmed (Fig. 3A,B).

The graft was retrieved from a 66-year-old woman considered to be not suitable for heart donation; this allowed us to harvest the liver with the right atrium en bloc (Fig. 4A).

In the recipient, the donor atrium was brought into the chest through the diaphragm for the atrium-atrium anastomosis (white circle in Fig. 4B). The anastomosis of the inferior vena cava, portal vein, hepatic artery, and bile duct was performed in a standard fashion.

During the procedure, hemodynamic parameters remained stable, the cold ischemia time was 7 hours, and 12 units of packed red blood cells were required. The postoperative course was uneventful, and the patient was discharged after 4 weeks.

We believe that the key point for the success of this procedure was the planning of liver and partial atrium transplantation with cardiopulmonary bypass. The use of the donor atrium ensured enough tissue to remove the recipient vena cava together with the portion of the atrium affected by the thrombus and easy accomplishment of the atrium-atrium anastomosis. The cardiopulmonary bypass and the partial atrium transplantation due to the atrium being affected by the thrombus, reported in other cases,^{4,5} permitted optimal management of the intraoperative hemodynamic conditions, counterbalancing the need for heparin infusion during extracorporeal circulation.

Abbreviation: TIPS, transjugular intrahepatic portosystemic shunt.

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Figure 1. Computed tomography scan shows the presence of a thrombus in the vena cava from the renal veins to the right atrium.

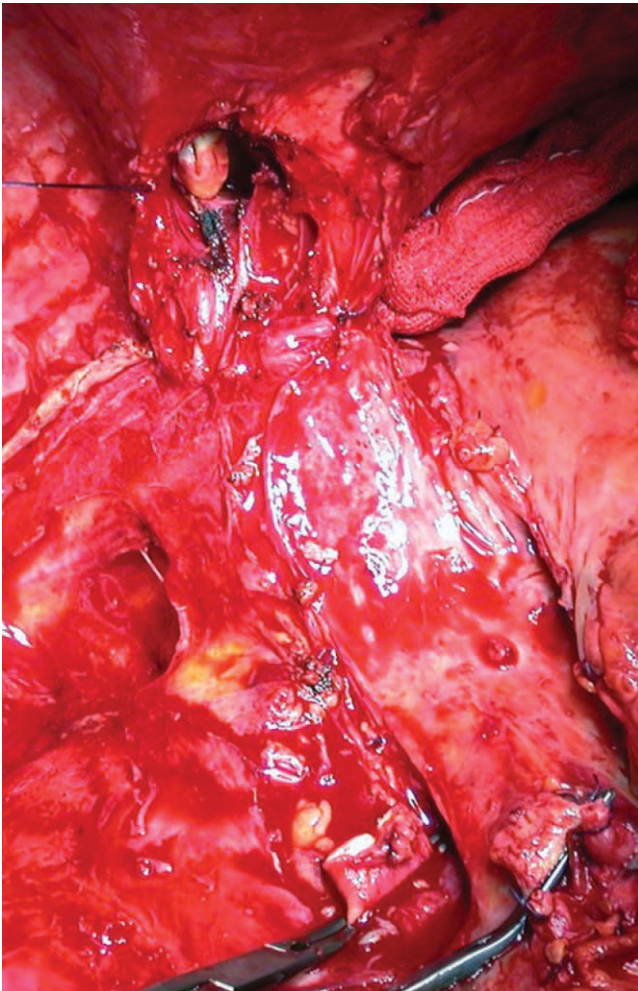


Figure 2. Recipient hepatectomy with the resection of the vena cava and the opening of the diaphragm through the central fibrous body.

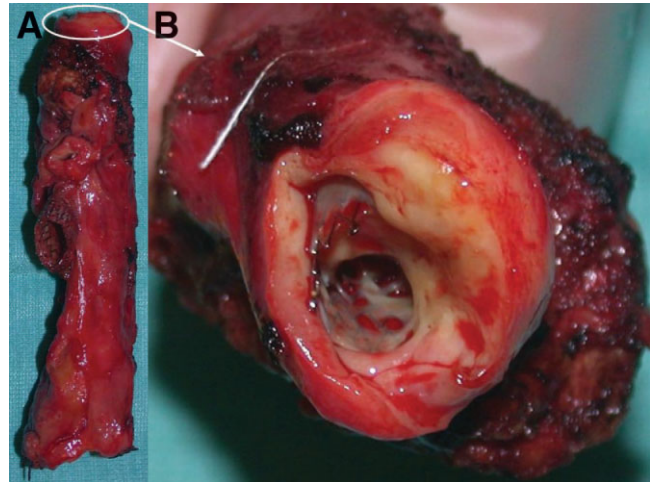


Figure 3. (A) Vertical view of the vena cava, which shows the TIPS in the suprahepatic vein, and (B) horizontal view, which shows the second TIPS up to the right atrium.

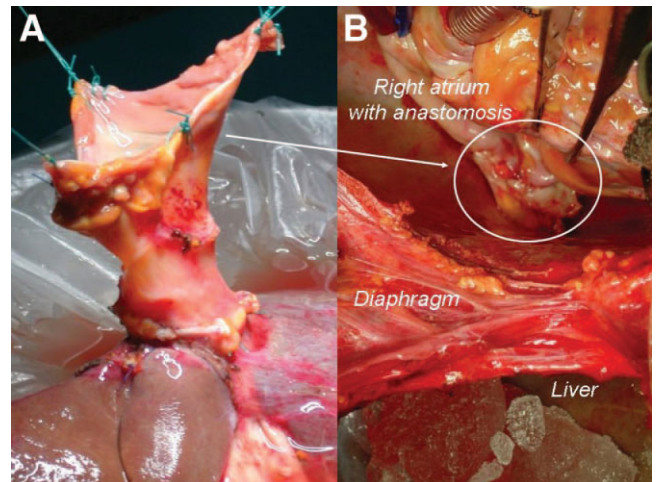


Figure 4. (A) Graft harvested with the liver and the right atrium en bloc and (B) donor atrium brought into the chest through the diaphragm for the atrium-atrium anastomosis (white circle).

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