
TRANSCARDIAC RELIEF OF BUDD - CHIARI SYNDROME WITH OBSTRUCTION OF THE INFERIOR VENA CAVA: A WORD OF CAUTION

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A 22 year old male patient with Behçet's disease was referred to surgery for progressive liver disease infiltrating the inferior vena cava (IVC) and the hepatic veins, presenting with evolving hepatic coma. Computed tomographic (CT) scanning revealed occlusion of the terminal portion of the IVC along with the hepatic veins. The patient was operated on an emergency basis, without inferior vena cavography. The patient was put on cardiopulmonary bypass (CPB) and the occlusion of the IVC and the hepatic veins was relieved through the right atrium in the beating heart. After an otherwise successful operation, removal of the IVC cannula, which was inserted via the femoral vein, caused a previously unsuspected IVC thrombus that was wedged around the cannula, to embolize to the pulmonary circulation. Pulmonary embolectomy did not permit the salvage of the patient. This bitter experience was caused by the undetected lower IVC thrombus, owing to reliance on the CT scanning alone. It is concluded that, transcatheter relief of Budd-Chiari syndrome with IVC obstruction is feasible and inferior venacavography should always be performed in these cases.

Key words: Hepatic vein thrombosis; vena cava inferior; Behçet's syndrome.

Occclusion of the hepatic veins along with the terminal portion of the inferior vena cava (IVC) is a rare entity and the overall prognosis for the patients is dismal and disappointing¹. Estrogen therapy, various hemathologic disorders, previous hepatic infections, cirrhosis, infiltrating diseases, and membranous obstructions are deemed responsible for the development of Budd-Chiari syndrome²⁻⁴. Not infrequently, no etiologic factors of associated disorders can be identified⁵. The goals of treatment of the Budd-Chiari

syndrome are relief of portal hypertension, relief of inferior vena cava (IVC) syndrome, if present, and preservation of hepatic function⁵. This report describes the surgical management of a case of Budd-Chiari syndrome associated with obstruction of the terminal portion of the IVC.

Case Report

A 22 year old male patient, known to have Behçet's disease was referred to surgery for IVC and hepatic vein occlusion with end-stage hepatic failure and massive ascites (Figure 1). The diagnosis was based on computed tomographic (CT) scanning, which revealed occlusion of the terminal IVC and the hepatic veins. The proximal IVC was reported to be patent. The diagnosis was deemed sufficient and the patient was taken into the operating room on an emergency basis, in an attempt to re-establish hepatic outflow and prevent the evolving hepatic coma.

A median sternotomy was performed and the skin incision was extended to the umbilicus, exposing the heart as well as the abdomen (Figure 2). Superior vena cava was cannulated in the usual manner, and the IVC was cannulated via the femoral vein, with a 30 cm long fenestrated venous cannula. The patient was put on cardiopulmonary bypass (CPB) with a membrane oxygenator, and the CPB circuit was primed with autologous ascites. Venous inflow into the CPB circuit from both venous cannulae were adequate. Arterial return was established via a cannula into the ascending aorta.

The liver, which was fibrotic and shrunked in appearance, was mobilized from its diaphragmatic attachments and the diaphragm was incised down to the IVC. Under normothermic CPB, the right atrium was opened in the beating heart. The inferior portion of the right atrium was filled with a soft infiltrating tissue, which was cleared accordingly. The right atrial incision was extended caudally to the IVC as far as possible while the liver was retracted manually. The IVC was occluded with a soft, white laminar infiltration. A thrombectomy catheter was used to relieve the vena caval infiltration. A right-angled clamp aided the insertion of the thrombectomy catheter into the he-

patic veins, which were cleared subsequently. Blood flow through the hepatic veins was observed, and air entered the CPB circuit via the IVC cannula, which was clamped in turn. The incision in the right atrium and the IVC was closed, and air was evacuated from the right heart chambers. Discontinuation of CPB was uneventful. As heparin neutralization was in progress, the IVC cannula was taken out and the femoral vein repaired. At this time a sudden increase in pulmonary artery pressure was noted with vigorous right ventricular contractions followed by hemodynamic deterioration. CPB was re-established immediately. It appeared that, fresh thrombus, which wedged around the IVC cannula was released following decannulation and caused massive pulmonary embolism. Although pulmonary embolectomy was performed and some fresh clot obtained from both of the pulmonary arteries, it was impossible to clean the distal pulmonary arterial tree completely. Several attempts to wean from CPB was unsuccessful.

Discussion

Occlusive lesions of the IVC may often complicate Budd-Chiari syndrome⁶. It is obvious that, operative procedures must be tailored to the etiology and pathology. The selection of the shunt-type operations are determined by IVC obstruction⁷⁻⁹. Most authors consider the mesoatrial shunt as the operation of choice for occlusion of the retrohepatic IVC and hepatic veins^{2,6,7,9}. The cava-atrial shunt is preferred for occlusion of the IVC and patent hepatic veins, membranotomy for IVC web, and the meso-caval shunt for intrahepatic venous occlusion^{6,8,10}. Recently transluminal angioplasty and/or expandable venous stents have been used for hepatic decompression in suitable cases^{3,5}. In the surgical management of acute Budd-Chiari syndrome complicated by IVC obstruction, a two stage approach was advocated by the Emory University group. This entailed initial hepatic decompression by suprahepatic mesoatrial shunt, with subsequent takedown of the mesoatrial shunt combined with conversion to a short infrahepatic portocaval shunt¹¹. Despite initial symptomatic improvement, this approach resulted in IVC stenosis on the long



Figure 1: The patient in the operating room. Note massive ascites and collateral formation.

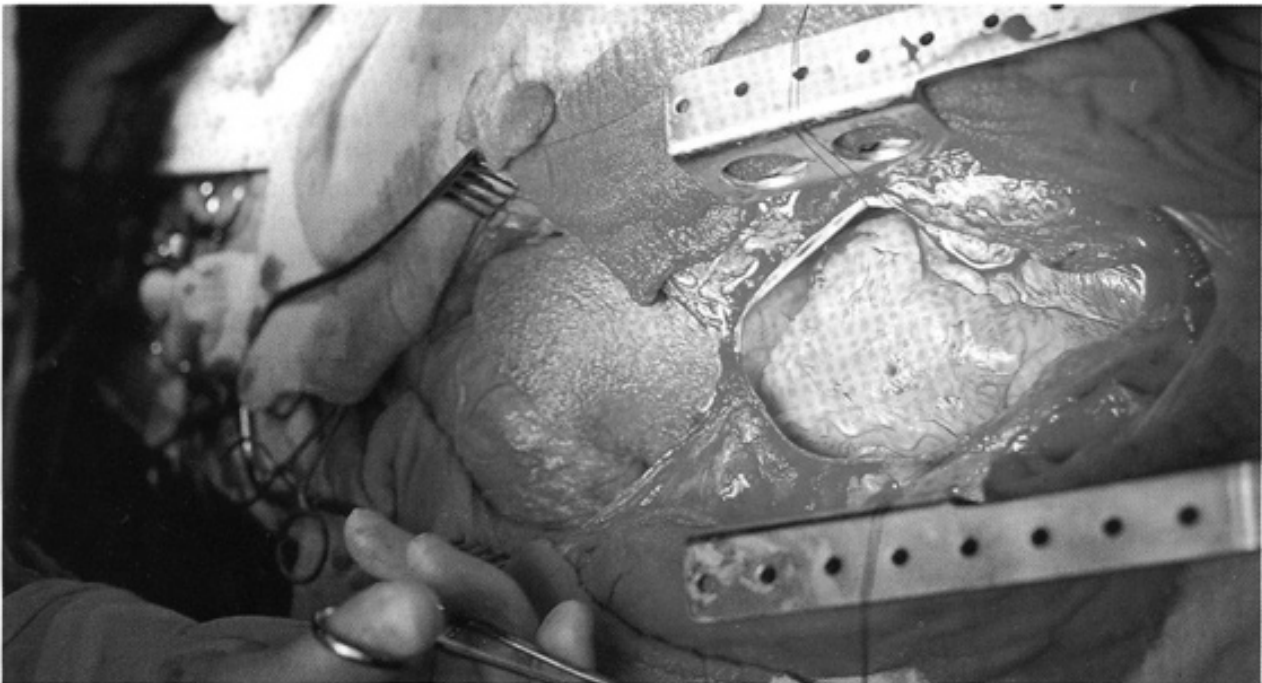


Figure 2: Surgical incision exposing the heart and the abdomen.

term¹¹. In general, mesoatrial shunt procedure is a demanding and disappointing procedure regarding early and long-term results². This has prompted us to adopt the transcatheter membranotomy technique, which is used successfully in the membranous obstruction of hepatic veins or IVC^{6,8,10}, to this case of Budd-Chiari syndrome with IVC obstruction. The technique proved to be successful, owing in part to the soft and laminar infiltration of Behçet's disease. Pulmonary embolism was not directly related to the procedure, which can be anticipated in this kind of surgery¹⁰.

In cases with Budd-Chiari syndrome, demonstration of the patency of the IVC is a major issue in planning the surgical approach^{7,12,13}. Detection of acute thrombosis of the IVC or hepatic veins by CT scanning can be troublesome to an inexperienced interpreter, lacking the knowledge of special CT features of acute IVC thrombosis¹⁴. Sophisticated techniques like magnetic resonance imaging or single photon emission computed tomography have evolved as valuable diagnostic tools in the recent years¹⁵⁻¹⁷. Likewise, duplex or color-flow doppler is being used as a primary imaging procedure^{4,12,13}. Despite this wide range of diagnostic methods, inferior venacavography should still be accepted as the gold standard in the demonstration of IVC occlusion^{2,7,13}.

Budd-Chiari syndrome can rarely be observed in patients with Behçet's disease^{18,19}. The case reported herein had end-stage liver disease with severe hepatocyte necrosis and massive fibrosis of the liver. In this situation the role of surgery is obviously palliative, and the aim is to achieve a degree of hepatic compensation which may permit liver transplantation as a lifesaving measure⁹.

In the light of this experience, it is concluded that, transcatheter relief of Budd-Chiari syndrome with IVC obstruction is feasible, and patency of the IVC should always be confirmed with inferior venacavography in cases where cannulation of the IVC is considered

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