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***Case report-Olgu sunumu***
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# Management of Budd Chiari Syndrome by liver transplantation: A case report

## *Budd Chiari Sendromu'nda karaciğer transplantasyon yönetimi: Bir olgu sunumu*

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**Abstract**

Budd-Chiari syndrome (obstruction of venous drainage of the liver) may be idiopathic, due to thrombosis of the hepatic veins in thrombogenic states or be due to extrinsic compression. It is rare, but life-threatening, if left untreated. For patients with end-stage liver disease, liver transplantation should be considered. Orthotopic liver transplantation (OLT) is associated with the potential for massive blood loss, necessitating rapid infusion of large quantities of blood products. In these patients, hemodynamic instability can be dramatically diminished by inferior vena cava cross-clamping during OLT. There are few reports about living donor transplantations in Budd-Chiari case. We present a case of 27-year-old man, with a diagnosis of end-stage liver disease secondary to Budd-Chiari syndrome and underwent living OLT. Anaesthesia management, and follow-up of patients affected by this condition represent a challenge for anesthesiologists. The aim of this report is to discuss the potential options in the intraoperative management of patients with Budd-Chiari syndrome, who will undergo living OLT.

**Keywords:** Budd-Chiari syndrome, orthotopic liver transplantation, anaesthesia management

**Özet**

Budd-Chiari sendromu (karaciğer venöz drenaj tıkanması) idiyopatik, hepatik venin trombojenik trombozu veya ekstrensek basısına bağlı gelişebilir. Nadir görülür, fakat tedavi edilmez ise yaşamı tehdit edicidir. Son dönem karaciğer hastalığı olan hastalar için, karaciğer transplantasyonu düşünülmelidir. Ortotopik karaciğer transplantasyonu (OKT), masif kan kaybı potansiyeli ve kan ürünlerinin büyük miktarlarda hızlı infüzyon gereksinimi ile ilişkilidir. Bu hastalarda OKT sırasında vena kava inferiora kros klempaj uygulanmasıyla hemodinamik instabilite önemli ölçüde azaltılabilir. Budd-Chiari ile birlikteliği olan karaciğer donör transplantasyonu olguları hakkında az sayıda rapor bulunmaktadır. Bu yayında, Budd-Chiari sendromuna sekonder olarak son dönem karaciğer hastalığı tanısı alan ve OKT yapılan 27 yaşındaki bir erkek hasta sunulmaktadır. Bu hastaların anestezi yönetimi ve takibi anesteziyolojistler için önemli bir sorun oluşturmaktadır. Bu raporun amacı Budd-Chiari sendromu ile birlikte OKT uygulanması planlanan hastaların intraoperatif yönetiminde potansiyel seçenekleri tartışmaktır.

**Anahtar sözcükler:** Budd-Chiari sendromu, ortotopik karaciğer transplantasyonu, anestezi yönetimi

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## Introduction

Liver transplantation is the most effective treatment for fulminant liver failure and cirrhosis. Orthotopic liver transplantation (OLT) is associated with the potential for massive blood loss, necessitating rapid infusion of large quantities of blood products [1]. This possibility justifies the use of invasive hemodynamic monitoring and large-bore venous access. Some patients may not tolerate the hemodynamic consequences of decreased venous return and cardiac output that can occur when the inferior vena cava (IVC) is cross-clamped [2].

Budd Chiari syndrome (BCS) is a rare disorder resulting from hepatic venous outflow tract with obstruction anywhere from the small hepatic veins to the suprahepatic inferior vena cava [3]. In these patients, hemodynamic instability can be dramatically diminished by inferior vena cava cross-clamping during OLT [4].

Herein, we present our anesthesia management in a patient with BCS and liver cirrhosis who underwent OLT. Potential options in the intraoperative management of these patients are discussed.

## Case report

A 27-year-old, 65 kg man, with a diagnosis of end-stage liver disease secondary to BCS was referred to our clinic for living OLT. Past medical history included recent renal failure (hepatorenal syndrome) requiring dialysis. The patient's Child- Pugh and Model for End-stage Liver Disease scores were 11 and 23, respectively (www.mayoclinic.org/meld/mayomodel6.html). Preoperative laboratory data showed a platelet count of 185.000 $mm^3$, hemoglobin level of 13,2 g/dL, activated partial thromboplastin time of 34.7, and international normalized ratio of 1.14 (0.9-1.4). Physical examination was significant for a massively distended abdomen secondary to ascites. Vital signs were as follows: heart rate was 120 beats/min, blood pressure was 85/55 mmHg, and an unlabored respiratory rate was 16 breaths/min. Initial intraoperative monitors included urine output; body temperature; electrocardiography; pulse oximetry, central venous pressure, and end-tidal carbon dioxide; and an arterial line for continuous blood pressure monitoring An induction of general anesthesia was accomplished with intravenous thiopental, 150 mg; fentanyl, 120 µg; and vecuronium, 10 mg. Endotracheal intubation was performed. Anesthesia was maintained with an 50% oxygen/50% air/isoflurane mixture. A double-lumen dialysis catheter was placed in the right internal jugular (RIJ) vein in the operating room. During the recipient surgery, supra and infra vena caval segments were clamped and the retrohepatic vena cava was opened to remove the thrombosed stent. The vena caval endothelium was completely disrupted with an extremely thin wall. Blood flow continued to the suprarenal part of the resected inferior vena cava. Clamping of the portal vein did not lead to significant hemodynamic changes or to bleeding from varices. The IVC was partially clamped at the confluence of the hepatic veins with only minor changes in hemodynamics. The intraoperative course was remarkable for a mild-to-moderate decrease (20% to 25%) in mean arterial blood pressure, which was treated easily with an infusion of epinephrine (50 to 150 ng/min/kg). Metabolic acidosis was corrected with sodium bicarbonate and hypocalcaemia was corrected with calcium gluconate during the operation. After reperfusion, the epinephrine was slowly discontinued, and the surgery was completed uneventfully. The total operative time was 10 hours and 45 minutes, total anaesthetic duration was 13 hours and 38 minutes, total blood loss was 2670 mL, total infused volume was 9750 mL (crystalloids 9000 mL, colloids 750 mL, albumin 6 U) and total blood transfusion was approximately 2500 mL (packed red blood cells 10 U, fresh frozen plasma 3 U). Urine output during anaesthesia was 2060 mL. The patient was transported to the surgical intensive care unit, and the endotracheal tube was removed 12 hours after the operation in the intensive care unit. The postoperative course was unremarkable.

## Discussion

BCS may be idiopathic or it may be due to the thrombosis of the hepatic veins in thrombogenic states or extrinsic compression, for example, due to neoplasia or hypercoagulability of the blood [5]. Flow obstruction results in portal hypertension and hepatocellular damage. Cirrhosis and portal hypertension eventually develop. Treatment for BCS should be in a stepwise manner. Medical supportive treatment with anticoagulants is applied as an initial therapy. When this approach fails, more-invasive therapeutic options should be considered [6]. For patients with end-stage liver disease, liver transplantation must be considered. Living-donor liver transplant might be a safe option, especially in countries where deceased donor grafts are of limited availability. There are few reports about living donor transplantations in BCS.

Preoperative investigation should be aimed at determining the extent of liver and other concomitant organ disease in addition to the effects of therapy (e.g. diuretics, anticoagulants, chemotherapy). Coagulation abnormalities should be corrected (fresh frozen plasma, platelets, vitamin K). In this case we administered 10 units of packed red blood cells and 3 units of fresh frozen plasma.

Anesthesia management of patients with BCS remains challenging. In a patient presenting for OLT for BCS, the anesthesiologist should be alert to the possibility of bleeding, or hypercoagulability [7]. If the obstruction develops over time, the patient may remain asymptomatic as adequate collateral circulation develops. This collateral circulation may not be adequate, however, to handle either rapid volume transfusion or increased venous flow secondary to venovenous bypass [8].

Patients with cirrhosis are usually in a hyperdynamic state with concomitant low peripheral vascular resistance. Hypotension can usually be controlled by careful titration of potent vasopressor agents, as needed, during OLT. Because the liver allograft was the right lobe, total clamping of the inferior vena cava was necessary during vascular reconstruction in our case. Total caval clamping decreases venous blood return to the heart, with a subsequent decrease in cardiac output and blood pressure. The next set of steps were to ensure that clamping of the confluence of the hepatic veins and portal vein would not lead to significant hemodynamic changes or to bleeding from varices. In our liver transplantation program, venovenous bypass has not been used in any living donor operation. The anhepatic phase in this case was approximately 1 hour. An acceptable mean blood pressure of 60 to 70 mmHg had to be maintained by increasing fluid requirements and correcting metabolic acidosis with sodium bicarbonate and hypocalcaemia with calcium gluconate.

Our patient tolerated the OLT, and the recovery was unremarkable. He was alive with the original graft and had satisfactory liver function. Therefore, efforts to maintain stable hemodynamics and to prevent hypotension are crucial in the management of a patient with an BCS undergoing OLT.

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