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Case report

Liver Transplantation as Treatment for Budd–Chiari Syndrome: Case Report

Álvaro Andrés Montenegro,[1*] Hernan Yair Florez-de-Moya,[2] Diego Mauricio Gómez,[3] Juan Manuel Rico-Juri,[4] Michael Zapata,[5] Jhonatan Urrego,[6] Luis Alfonso Bustamante-Cristancho.[7]

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[1] Specialist in Emergency Medicine; Specialist in Critical Care Medicine and Intensive Care; Specialist in Epidemiology. Clínica Imbanaco. Cali, Colombia.
[2] Emergency Medicine Resident, Pontificia Universidad Javeriana (PUJ) Cali. Clínica Imbanaco. Cali, Colombia.
[3] Gastroenterology and Hepatology, Liver Transplant Group. Clínica Imbanaco. Cali, Colombia.
[4] Hepatobiliary and Transplant Surgeon. Clínica Imbanaco. Cali, Colombia.
[5] General Surgeon. Clínica Imbanaco. Cali, Colombia.
[6] Specialist in Intensive Care. Adult Intensivist at Clínica Imbanaco. Cali, Colombia.
[7] Emergency Physician, Intensivist, Radiologist. ICU Coordinator, Imbanaco. Clínica Imbanaco. Cali, Colombia.

***Correspondence**: Álvaro Andrés Montenegro.
alvaro.montenegro@javeriana.edu.co

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Abstract

Background: Polycythemia vera is a myeloproliferative neoplasm that leads to hyperviscosity, blood stasis, and a high risk of thrombotic complications. When these events affect the hepatic veins, they give rise to Budd–Chiari syndrome, characterized by elevated portal capillary hydrostatic pressure, predisposing patients to ascites, variceal bleeding, and hepatic dysfunction. Liver transplantation is considered in two principal clinical scenarios: fulminant hepatic failure and progressive cirrhosis, particularly when TIPS is not feasible. **Case summary**: We report the case of a 40-year-old woman with polycythemia vera and Budd–Chiari syndrome who, despite successful recanalization of the hepatic veins through anticoagulation, experienced clinical deterioration with progressive portal hypertension and refractory ascites. Her severity score and clinical decline (Rotterdam index 2.7 = class III, poor 3-month prognosis) supported the decision to proceed with a deceased-donor liver transplant, which was successfully performed. Sequential use of antiplatelet therapy, hydroxyurea, and anticoagulation prevented thrombotic complications without inducing bleeding. Outpatient follow-up showed no clinical signs of portal hypertension, including ascites or variceal bleeding. **Conclusion:** Budd–Chiari syndrome secondary to myeloproliferative disorders is uncommon, and its management must be individualized according to disease stage, comorbidities, and institutional expertise to achieve optimal patient outcomes.

Keywords

Polycythemia vera, Budd–Chiari syndrome, liver transplantation, acute myeloid leukemia secondary to myelodysplastic syndrome.

INTRODUCTION

Polycythemia vera is considered a myeloproliferative neoplasm characterized by the clonal expansion of an abnormal hematopoietic stem cell, resulting in elevated blood cell counts—predominantly red blood cells—which leads to hyperviscosity and blood stasis and, consequently, a predisposition to thrombotic complications[(1)] affecting both the arterial and venous vascular systems[(2)]. When the thrombotic event involves the inferior vena cava or the hepatic veins, it constitutes Budd-Chiari syndrome, which is characterized by a hemodynamic disturbance secondary to increased portal capillary hydrostatic pressure. This results in sinusoidal dilatation and leakage of fluid into the interstitial space, with a consequent increase in portal pressure and a reduction in hepatic perfusion, leading to cellular hypoxic injury[(3)].

Budd-Chiari syndrome is one of the indications for liver transplantation, usually associated with two forms

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of disease presentation: fulminant hepatitis or cases of progressive cirrhosis, particularly when transjugular intrahepatic portosystemic shunt (TIPS) placement is not feasible[4]. We present a case of cirrhosis due to Budd-Chiari syndrome in a patient who was not a candidate for TIPS because of recanalization of the hepatic veins resulting from anticoagulation therapy.

CLINICAL CASE

We report the case of a 40-year-old female patient with no significant past medical history, who had been diagnosed with Janus kinase 2 (JAK2)-positive polycythemia vera 12 years earlier. Four years prior to the current clinical presentation, partial thrombosis of the portal vein and hepatic veins was documented (**Figure 1**). She was therefore treated with anticoagulation using warfarin (2.5 mg five days per week alternating with 5 mg two days per week), without achieving adequate control of the international normalized ratio (INR) during follow-up. Anticoagulation was subsequently switched to apixaban, in combination with hydroxyurea (500 mg daily). Ten months later, a repeat abdominal computed tomography scan demonstrated recanalization of the portal vein thrombosis and a high suspicion of recanalization of the hepatic vein thrombosis, which was later confirmed by cavography.

Despite continued anticoagulation and evidence of recanalization of the hepatic vein thrombosis, over the last four months the patient progressively developed symptoms consistent with large-volume ascites (greater than 10 liters), requiring frequent drainage (approximately every two weeks). This was accompanied by marked weight loss (approximately 15 kg over the same period) and an episode of hepatic encephalopathy classified as grade III according to the West Haven criteria, which required hospitalization in the intensive care unit, in addition to several grade I–II episodes managed on an outpatient basis. During hepatology follow-up, liver cirrhosis was diagnosed based on the clinical presentation, laboratory findings (**Table 1**), and imaging studies (transient elastography showing 14.16 kPa, compatible with cirrhosis; reference >11.9 kPa). Other etiologies were ruled out, including alcoholic liver disease, metabolic dysfunction-associated fatty liver disease, and autoimmune or infectious liver disease (negative antinuclear and anti-smooth muscle antibodies, negative hepatitis B surface antigen, and negative hepatitis C antibodies). Given advanced liver dysfunction associated with encephalopathy and a Rotterdam score of 2.7, the patient was classified as class III (poor three-month prognosis). Evaluation for liver transplantation was therefore pursued, and the possibility of TIPS placement was ruled out. In a multidisciplinary meeting, she was listed for transplantation with a Child-Pugh class B (9 points) and a MELD-Na score of 16.

Three months after being placed on the waiting list, a graft from a deceased donor became available. Intraoperative findings revealed changes consistent with severe portal hypertension, including the presence of splenorenal and retroperitoneal shunts and recanalization of the umbilical veins, as well as hepatomegaly, with a liver weight of 2,780 g. Orthotopic liver transplantation was performed without recorded complications (total ischemia time of 10 hours and 29 minutes; warm ischemia time of 35 minutes).

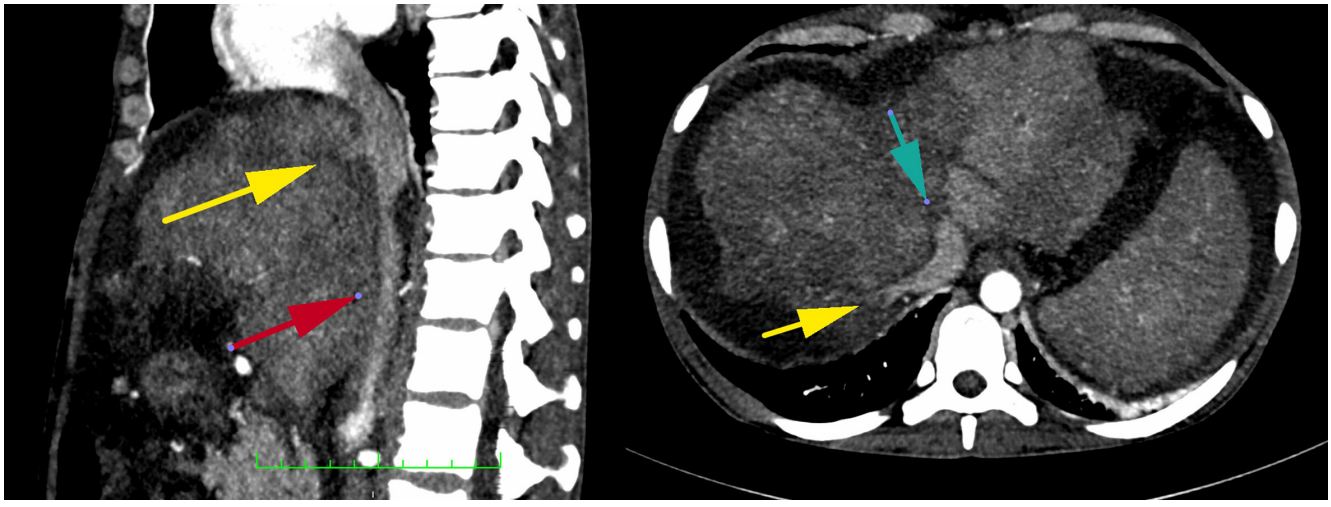


Figure 1. Abdominal CT Angiography Acquired in the Arterial Phase. The right hepatic vein shows flow interruption, suggesting chronic-appearing occlusion (yellow arrow). The middle hepatic vein also shows flow interruption, suggesting chronic-appearing occlusion (green arrow). The retrohepatic inferior vena cava shows reduced caliber without thrombosis or occlusion (red arrow). Author's file.

Table 1. Paraclinical Follow-up and Portal Doppler after Transplantation

	Outpatient Follow-up	Day 1	Day 3	Day 5
ALT (U/L)	22	623	366	230
AST (U/L)	17	1087	159	51
INR	1.35	2.83	1.42	-
Total bilirubin (mg/dL)	0.30	2.89	0.58	0.56
Direct bilirubin (mg/dL)	0.15	2.45	0.40	0.3
Hemoglobin (g/dL)	11	10.7	7.6	7.7
Creatinine (mg/dL)	0.66	0.65	0.82	0.85
Tacrolimus levels (ng/mL)	NA	-	-	5.0
Portal Doppler hepatic artery RI	NA	0.62 (12 h)	0.6 (24 h)	0.67 (48 h)

ALT: alanine aminotransferase; AST: aspartate aminotransferase; INR: international normalized ratio; RI: resistance index; NA: not applicable. Author's own research.

Postoperatively, the patient was transferred to the intensive care unit with invasive mechanical ventilatory support and vasopressors. She subsequently showed satisfactory evolution, with withdrawal of ventilatory support and complete discontinuation of vasopressors after 48 hours in the unit. Immunosuppression was administered according to the institutional protocol (tapering doses of methylprednisolone and tacrolimus). During the first 24 hours, antiplatelet therapy with acetylsalicylic acid (ASA) (100 mg/day) and hydroxyurea 500 mg/day was indicated. Enoxaparin (40 mg subcutaneously once daily) was initiated within the first 48 hours, with no evidence of bleeding complications.

After completing 48 hours in the intensive care unit, transfer to the general ward was indicated. Her clinical course was satisfactory, with improvement in cytolysis, cholestasis, and liver function parameters (**Table 1**). Given the high thromboembolic risk, serial portal Doppler evaluations were performed at 12, 24, and 48 hours in the unit, all showing normal findings (**Table 1**). She was discharged on postoperative day five with instructions for outpatient medical management, including prednisolone 20 mg/day, extended-release tacrolimus 2 mg/day, hydroxyurea 500 mg/day, ASA 100 mg/day, trimethoprim/sulfamethoxazole 160/800 mg three times per week, enoxaparin 40 mg subcutaneously once daily for one month, fluconazole 200 mg/day for 21 days, and furosemide 40 mg/day.

At the two-month follow-up visit, the patient showed adequate clinical progression, with no new episodes of ascites, variceal bleeding, or hepatic encephalopathy. At the most recent follow-up, a weight gain of 1 kg compared with her previous weight was documented.

DISCUSSION

Polycythemia vera is considered a myeloproliferative neoplasm characterized by the clonal expansion of an abnormal hematopoietic stem cell, resulting in elevated blood cell counts—predominantly red blood cells—most commonly due to different mutations in the JAK2 tyrosine kinase gene. Approximately 97% of these mutations correspond to JAK2 V617F, which results from a somatic guanine-to-thymine mutation involving exon 14 of the JAK2 gene. This leads to a nucleotide change at position 1849 and the substitution of valine by phenylalanine at codon 617[5,6]. The pathophysiological consequence is stasis due to hyperviscosity and an increased risk of thrombotic events associated with blood stasis. Thrombotic events usually occur in the venous system and are more prevalent in women, whereas arterial thrombosis is less frequent[2]. In the present case, the diagnosis of polycythemia vera was supported by the course of erythrocytosis, thrombotic phenomena, and the presence of a JAK2 mutation; accordingly, the patient was being followed on an outpatient basis by hematology.

Budd-Chiari syndrome is defined as hepatic venous outflow obstruction, leading to hypotrophy or atrophy of the affected hepatic segment and hypertrophy of the remaining segments, with symptoms that depend on the extent of the obstructive involvement. The consequence of this obstruc-

tion is a hemodynamic disturbance caused by increased portal capillary hydrostatic pressure, which alters vascular pressure gradients. These hemodynamic changes translate into sinusoidal dilatation and leakage of fluid into the interstitial space, passing through the hepatic capsule when lymphatic drainage capacity is exceeded, with a consequent increase in portal pressure and a reduction in hepatic perfusion, resulting in cellular hypoxic injury[3]. In the present case, thrombosis of the hepatic veins established the diagnosis of Budd-Chiari syndrome, and the patient also developed symptoms related to clinically evident portal hypertension due to the presence of large-volume ascites.

Budd-Chiari syndrome is one of the indications for liver transplantation and is usually associated with two forms of disease presentation. The first is fulminant hepatitis with secondary cirrhosis, which results from acute obstruction of hepatic venous outflow, leading to hepatic congestion and rapid-onset dysfunction. The second presentation is subacute, consisting of gradually developing liver injury, likely because some patients develop intrahepatic and portosystemic collaterals and may therefore remain asymptomatic for prolonged periods[4]. When symptoms of portal hypertension develop, clinical manifestations such as persistent ascites, esophageal varices, or variceal bleeding may occur[7]. The reported case fits the second form of presentation, given its chronic course and the presence of portal hypertension associated with portosystemic collaterals. In this patient, recanalization of the hepatic veins was confirmed by cavography, likely related to chronic anticoagulation; however, the clinical course suggests progressive disease characterized by persistent ascites and other complications, such as spontaneous bacterial peritonitis or hepatic encephalopathy. This indicates that the patient developed liver cirrhosis, probably secondary to Budd-Chiari syndrome, which did not resolve despite recanalization of the hepatic veins.

Recent publications suggest that the prognosis of patients undergoing liver transplantation for Budd-Chiari syndrome is comparable to that of other etiologies[8], provided that anticoagulant therapy is maintained postoperatively[9]. However, in patients with myelodysplastic syndromes, outcomes appear to be somewhat less favorable in terms of survival[10]. In this regard, one study followed 14 patients after liver transplantation indicated for Budd-Chiari syndrome and reported three postoperative deaths due to primary graft nonfunction, heart failure, and intra-abdominal sepsis; another patient died from recurrence of Budd-Chiari syndrome 19 months after transplantation. Early complications described included gastrointestinal bleeding, cholangitis and mechanical ileus, fistula and intra-abdominal abscess, pleural effusion, and surgical site infection. Three patients required intervention with endoscopic retrograde cholangiopancreatography and stent placement for biliary leaks. Nevertheless, one- and five-year survival rates were 87% and 71%, respectively[11], making liver transplantation an attractive therapeutic alternative for patients with fulminant liver failure and for those with portal hypertension refractory to medical management, particularly when TIPS is not feasible.

For the reported case, the indication for liver transplantation without TIPS was based on three main reasons. First, follow-up imaging studies demonstrated recanalization of the hepatic veins, suggesting that the clinical deterioration was most likely related to liver cirrhosis as a consequence of Budd-Chiari syndrome. Second, the clinical course was associated with hepatic encephalopathy, which could worsen with the use of TIPS. Finally, the third argument was related to the deterioration in quality of life in a young patient, in whom recurrent episodes of large-volume ascites and an episode of encephalopathy translate into increased mortality. It should be noted that, in this context, liver transplantation carries a higher risk of hepatic vascular thrombosis (arterial, venous, and portal) compared with liver transplantation for other etiologies; likewise, survival is lower than that observed for other indications for liver transplantation. Despite this, in several scenarios it becomes the only therapeutic alternative with acceptable survival rates. Recent literature has shown that 10-year survival after liver transplantation is not modified by prior TIPS placement or the absence thereof[12], allowing us to infer that appropriate patient selection based on disease assessment, comorbidities, and the experience of the treating center is critical when choosing the optimal therapy.

In the country, few studies have been published with follow-up of patients with Budd-Chiari syndrome. The first study characterized a cohort of 31 patients in the city of Bogotá, identifying a prothrombotic factor as the main etiology and, secondarily, myelodysplastic syndrome. In that cohort, 80% of patients received anticoagulation, 19% underwent TIPS, and 16% underwent liver transplantation. The reported mortality was 25.8%, with a mean survival of 337 days, compared with the death of two patients who underwent liver transplantation (12.5%)[13]. A second patient cohort was published in the city of Medellín, describing a subacute presentation of this condition in 35 patients; only 20 patients received recanalization treatment by different methods, and of these, only 20% underwent orthotopic liver transplantation[14]. In the present case report, survival of 12 months has been observed at the time of reporting, in the absence of complications such as portal hypertension, hepatic encephalopathy, or spontaneous bacterial peritonitis. Likewise, no complications such as vascular thrombosis have been recorded during the postoperative follow-up period.

In the post-liver transplantation follow-up of patients with polycythemia vera and Budd-Chiari syndrome, efforts focus on preventing recurrence of thrombotic events through the use of anticoagulants or antiplatelet agents. Lifelong anticoagulation with low-molecular-weight heparin has been considered, with warfarin as an alternative, adjusted to maintain an INR of 2–3[3]. However, during a mean follow-up of 28.2 months in 16 patients, several thrombotic events were reported (three in the portal vein, one in the hepatic artery, and one in the axillary vein), along with an even more concerning rate of hemorrhagic complications of 44%[15]. Additionally, reports have described management with hydroxyurea and aspirin instead of anticoagulant therapy in patients with documented myelodysplastic disorders, based on the theoretical cause of qualitative and quantitative platelet abnormalities. In these reports, aspirin was administered at a dose of 325 mg/day and hydroxyurea at doses ranging from 500 to 1,500 mg/day, initiated four to seven days after transplantation, with excellent long-term outcomes: 90% of treated patients did not develop new thrombotic events during up to 59.9 months of follow-up, and a 15-year survival rate of 73% was reported[15,16]. In the present case, early initiation of ASA, thromboprophylaxis with low-molecular-weight heparins, and hydroxyurea was chosen, with no documented recurrence of thrombotic events during the 12 months of postoperative follow-up, in the absence of bleeding and with a satisfactory clinical course that allowed hospital discharge on the fifth postoperative day after liver transplantation.

CONCLUSION

Polycythemia vera is associated with thrombotic phenomena, occasionally involving the hepatic veins and resulting in Budd-Chiari syndrome. This rare entity is associated with portal hypertension and complications such as variceal bleeding and hepatic encephalopathy. Treatment should be individualized according to disease progression, comorbidities, and the experience of the treating center. We present a case of liver cirrhosis due to Budd-Chiari syndrome successfully treated with liver transplantation.

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