Portal Steal Syndrome After Full-Size Deceased Donor Liver Transplantation

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ABSTRACT

Successful liver transplantation typically results in an immediate decrease in intrahepatic resistance accompanied by an initial increased hepatopedal portal flow. Within a short period of time, the portal hypertension resolves and the variceal shunts involute. However, in situations in which intrahepatic vascular resistance to venous flow remains elevated, significant hepatofugal portal flow may continue through persistent mesenteric shunts. This situation, portal steal, can result in decreased perfusion of the liver graft leading to graft dysfunction, failure, and potentially recipient death. This report details a case and the surrounding literature to highlight appropriate diagnosis and management in these patients.

INTRODUCTION

Shunting of blood flow through spontaneous portosystemtic connections commonly develops in patients with portal hypertension and can be identified in up to 19% of patients awaiting liver transplantation.[1] With progressive cirrhosis and the associated increased resistance to intrahepatic venous blood flow, mesenteric venous flow becomes hepatofugal through the splenic and/or left renal and coronary veins.[2-5] If this pathologic flow pattern is not identified and appropriately managed, patients undergoing orthotopic liver transplantation are at increased risk of morbidity and mortality. Successful liver transplantation typically results in an immediate decrease in intrahepatic resistance accompanied by an initial increased hepatopedal portal flow. Within a short time period, the portal hypertension resolves and the variceal shunts involute. However, in situations in which intrahepatic vascular resistance to venous flow remains elevated, significant hepatofugal portal flow may continue through persistent mesenteric shunts.[3-5] This situation, portal steal, can result in decreased perfusion of the liver graft leading to graft dysfunction, failure, and potentially recipient death.[5]

Portal vein steal syndrome has been described in situations of small liver volumes (live donor grafts and reduced-size or split liver grafts).[2] Here we report the diagnosis, management, and outcome of a patient with this syndrome who was diagnosed after a full-size deceased donor liver transplantation, review the current literature, and discuss best practice guidelines for prevention, evaluation, and management of this condition.

CASE REPORT

A 51-year-old obese woman with end-stage liver disease secondary to alcohol-induced cirrhosis presented for evaluation. Her past medical history included portal hypertension, esophageal varices, ascites, and hepatic encephalopathy. She underwent liver transplantation with a physiologic Model for End-Stage Liver Disease (MELD) score of 31 using a donation after cardiac death liver graft. The transplantation technique included portosystemic veno-venous bypass and cava replacement (Figure 1A-B). She remained hemodynamically stable throughout the transplant. The graft remained soft with excellent portal vein and hepatic artery perfusion by gross examination.

Immediate postoperative bedside duplex Doppler ultrasonography demonstrated bidirectional flow in the right and left portal veins and normal hepatic artery flow characteristics. Post-transplant day 1 (POD 1), laboratory evaluation demonstrated significant liver graft dysfunction with persistent elevation in alanine transaminase (ALT) and aspartate transaminase (AST) (Table 1). She was returned emergently to the operating room (OR) for evaluation of the graft and liver biopsy. Intraoperative duplex

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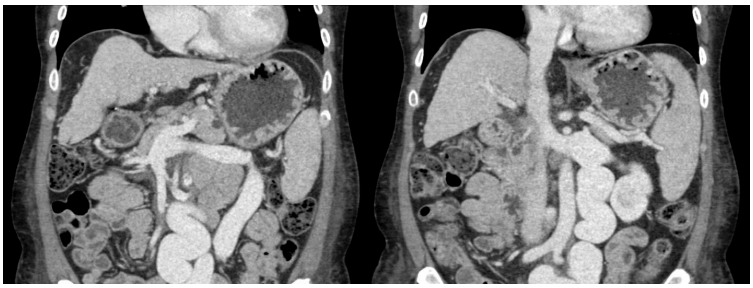
Figure 1A-B. Computed Tomography (CT) Scan Prior to Liver Transplantation



CT scan prior to liver transplantation demonstrating a large inferior mesenteric vein and splenorenal shunt.

Table 1. AST and ALT Lab Value Trends

Date	Pre-op 10/30	Post-transplant 10/31	POD 1 After Ligation of IMV 11/1	11/2	11/3	11/4	11/5	11/6	11/7	11/8
AST (10 - 32 U/L)	98	6671	3656	1426	632	226	104	55	40	34
ALT (8 - 33 U/L)	73	2844	2486	1413	939	554	373	244	181	135

Abbreviations: POD, postoperative day; IMV, inferior mesenteric vein; AST, aspartate transaminase; ALT, alanine transaminase.

Doppler ultrasound demonstrated hepatofugal and bidirectional flow in the portal vein. Portal venogram through a catheter placed in the proximal inferior mesenteric vein demonstrated persistent retrograde flow through the inferior mesenteric vein (IMV) and splenic vein. The portosystemic shunt was interrupted by ligation of the IMV. Post ligation intraoperative venogram demonstrated normal intrahepatic portal flow with no evidence of retrograde flow in the IMV or splenic vein (Figure 2B). She tolerated the procedure well, with normalization of liver enzyme values postoperatively (Table 1) and was eventually discharged to home with excellent graft function.

DISCUSSION

Portal steal syndrome results from persistent diversion of portal flow away from the liver through meso-systemic collaterals after liver transplantation. In patients who require transplant, the potential for portal steal syndrome should be identified prior to liver transplantation so that large and/or hemodynamically significant collateral vessels may be interrupted (ligation or coil embolization) during the initial transplant operation. Two distinct but occasionally coincidental issues related to this portal steal phenomenon can increase its likelihood, namely ischemia/reperfusion injury causing damage to the liver with increased resistance to flow or large shunts in the recipient causing diversion of portal blood flow away from the liver. Postoperative occurrence because of persistence of a large spontaneous shunt can result in graft failure due to reduced and/or reversed portal perfusion and a reduction in hepatotrophic factors.[5] Table 2 depicts risk factors for portal steal syndrome.[6-9] Persistent shunts can be difficult to identify and may require a multimodal approach during the intraoperative period to ensure complete ligation and adequate graft flow.[10] Prophylactic intraoperative exploration, evaluation, and ligation of large collateral splenorenal shunts (> 10 mm) appears to be the most effective way to prevent portal steal syndrome.[5-10]

Splenorenal shunts larger than 10 mm at their transition into the left renal vein have a high likelihood of portal steal and require operative intervention to ensure adequate liver transplant flow.[5] Lee and colleagues reviewed 44 cirrhotic patients with large spontaneous splenorenal shunts (> 10 mm in diameter).[5] All patients underwent living donor liver transplant with ligation of the left renal vein at the time of transplantation. Although portal flow increased after ligation of the left renal vein, 9.1% of patients demonstrated an elevated serum creatinine level after ligation.[5] The authors concluded that preemptive ligation of the left renal vein at the time of liver transplantation prevented a portal steal phenomenon.[5] Avoiding graft hyperperfusion by excessive portal hypertension is equally as important as preventing portal steal through large spontaneous collaterals.[5] Horrow et al[3] described a large spontaneous splenorenal shunt following orthotopic liver transplant, in an allograft with 10% macrosteatosis and a cold ischemic time of 9 hours and 26 minutes.[3] The routine sonography on POD 1 showed a patent anastomosis, but there was notable low portal venous flow. On POD 2, their patient had elevated liver function tests and a repeat sonography demonstrated bidirectional flow in the portal vein similar to our patient. The patient underwent reoperative surgery and an intraoperative sonogram showed the splenorenal shunt with hepatofugal flow, confirming steal from the liver.[3] After ligation of the splenorenal shunt, intraoperative sonography showed marked improvement in portal flow with velocities of 15-20 cm/sec as with our patient. Liver biopsy showed ischemia-reperfusion injury. The patient left the operating room, but died later that day.[3]

Vessels smaller than 10 mm may involute and hence do

not always require surgical intervention beyond liver transplantation.[5] Kim et al[7] described 19 patients with venous variceals following liver transplantation, of which 13 patients underwent intraoperative transvenous embolization resulting in 100% improvement in portal vein inflow.[7] Of their cohort, 6 patients underwent percutaneous transvenous embolization, but 33% showed technical failure and persistent portal steal. At 8-month follow-up, varices involuted in 13 patients, decreased in caliber 4, and remained unchanged in 2 patients.

Portal hemodynamics change dramatically following liver transplantation, and multiple studies have detailed the impact that shunting has on this dynamic. Jiang et al[8] examined differences in portal hemodynamics between whole liver transplantation and living donor liver transplantation and noted that the portal venous flow in patients with portal hypertension showed a high perfusion state after living donor liver transplant (LDLT) and, in contrast to the whole liver transplantation, portal venous pressure elevation after LDLT delaying the time necessary to close the collateral circulation.[8] Sainz-Barriga et al[9] prospectively evaluated intraoperative portal hemodynamics of 103 whole and partial liver transplants and found that portal vein flow and hepatic artery flow did not immediately return to normal values after liver transplantation.[9] Clinical outcomes of patients who underwent management of large collaterals to manage portal steal syndrome are summarized in Table 3.[3,5,7,10,11]

Aucejo et al[12] in an analysis of liver transplant recipients showed the utility of preoperative flow measurement by computed tomography (CT) for identifying potentially problematic shunts.[12] However, this may not find all collaterals as some shunts tend to be underperfused and tortuous in nature during the preoperative assessment. Kim et al[7] showed that intraoperative venography can significantly improve outcomes by quickly identifying newly engorged shunts, while still in the OR allowing early ligation before a threat to the graft occurs. Furthermore, intraoperative venography is not constrained by the tortuosity of the vessel.

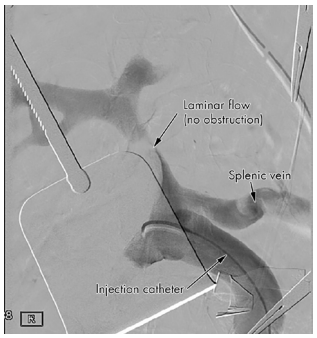
Smaller collaterals may become troublesome in the postoperative period as they can be missed by traditional imaging modalities and may mature into larger vessels during the postoperative period. Moon et al[10] showed that intraoperative portofluoroscopy as an adjunct to intraoperative ultrasonography (IOUS) and visual inspection can dramatically improve identification of potentially problematic collaterals and assist in the ligation of collaterals that would be missed otherwise. Intraoperative portofluoroscopy has the added benefit of providing accurate measurement of portal flow.[10]

Judicious ligation of shunts is necessary and requires sound clinical judgment as overly aggressive ligation can overwhelm the portal system. Initial studies suggest both the safety and efficacy of this practice to reduce graft failure rates from poor portal perfusion, re-operative intervention, and the need for postoperative angiography.

Portal steal syndrome is most common among patients with preoperative portal vein hypertension such as those with cirrhosis. While all liver transplant recipients may be affected, it appears to impact those with LDLT the most. Specific signs and symptoms of posttransplant portal vein steal syndrome are poorly reported, but tend to mimic signs and symptoms of acute rejection including poor clinical course, elevated liver function tests (LFT), and elevated total bilirubin. Doppler ultrasound may show bidirectional or hepatopedal flow in the portal vein.

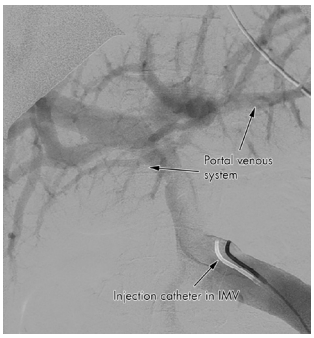
Identifying the potential for portal steal prior to liver transplantation is essential for patients undergoing transplant. We suggest utilizing imaging modalities such as magnetic resonance

Figure 2A. Preligation Intraoperative Mesenteric Venogram



Preligation intraoperative mesenteric venogram demonstrating retrograde flow in the splenic and inferior mesenteric veins. Laminar flow characteristics are seen within the inferior mesenteric vein.

Figure 2B. Postligation Intraoperative Mesenteric Venogram



Postligation intraoperative mesenteric venogram demonstrating prompt hepatopedal flow in both the right and left branches. The injection catheter within the inferior mesenteric vein is seen distal to the ligation site.

Table 2. Risk Factors for Recipient Portal Steal Syndrome

Portal hypertension: Large varices/shunts, chronic liver failure.
Macrosteatosis.
Low liver mass: Living donor liver transplant, split livers.
Donation after cardiac death: Prolonged warm ischemia time.
Receiving a living donor liver transplant.

Table 3. Summary of Publications Detailing Patients with Portal Steal Syndrome

Author	Mean Age	Study Size	Presenting Symptoms	POD No.	Treatment	Graft Saved	Mortality
Lee (2006)[5]	51 (26-64)	44	PV diameter 9.2 +/- 3.3, reduced PV flow, absent DUS signal in 7, hepatofugal DUS signal in 3, hepatopetal flow in all grafts, elevated T bili in all grafts.	1 day - 3 months	Ligation of LRV	43 alive at 17 mo follow-up	1/44
Moon (2007)[10]	48.8 (+/- 7.5)	5	Insufficient portal inflow and PV stenosis on DUS. MDCT showed stenosis, congestion, of L PV anastomosis. Elevated T bili.	3 Intra-operative 2 POD 4-5	Intraoperative Cine-portogram and ligation of patent collaterals	5 Saved	0/5
Kim (2009)[7]	46 (+/- 14.0)	19	Not reported.	46.4	Percutaneous transvenous embolization (6), intraoperative transvenous embolization (13).	17 Saved 2 Lost	3/19
Shirouzu (2009)[11]	33	1	Acute rejection, jaundice, coagulopathy, massive ascites, cholestasis, encephalopathy.	10	Proximal ligation of collateral splenorenal shunt w/o splenectomy or splenic artery ligation.	Saved	0/1
Horrow (2010)[3]	51	1	DUS showing bidirectional flow, deteriorating clinical cond., elevated LFTs.	2	Ligation of SSRS	Saved	1/1

Abbreviations: POD, postoperative day; PV, portal vein; DUS, Doppler ultrasonography; MDCT, multidetector row computerized tomography; LFTs, liver function tests.

imaging (MRI) and multi-phase CT to identify collaterals that may cause reversal of flow. Angiography should be the gold standard as this will also specifically identify those splenorenal shunts that are >10 mm at their transition into the left renal vein and hence have a high likelihood of portal steal. Once these have been identified, ligation or coil embolization during the initial transplant operation should be performed. Overall, we recommend prophylactic intraoperative exploration and ligation of these collateral splenorenal shunts (>10 mm) as this appears to be the most effective way to prevent portal steal syndrome.[5-10]

CONCLUSION

The differential diagnosis of immediate liver graft dysfunction should include a high index of suspicion for postoperative portal steal syndrome. The most effective therapy to avoid this complication may be the prophylactic ligation of potentially problematic shunts. This requires a multimodal approach and sound surgical judgment. The keys to successful outcomes postoperatively are having a high index of suspicion for portal vein steal syndrome to enable early recognition, regular ultrasound screening, and prompt institution of surgical therapy in order to salvage patients with portal vein steal syndrome.

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