

CASE REPORT

Portal Vein Recanalization Transjugular Intrahepatic Portosystemic Shunt for Noncirrhotic Chronic Portal Vein Thrombosis Leading to Successful Pregnancy: Case Report



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Portal vein recanalization transjugular intrahepatic portosystemic shunt (PVR-TIPS) is a safe and effective procedure for decompression of portal hypertension (PH). In this short case series, 2 women with chronic non-cirrhotic portal vein thrombosis were treated with PVR-TIPS. Both patients hoped to conceive. Without treatment for their PH, their pregnancies posed a significant risk of life-threatening variceal bleeding. Both patients tolerated the procedure well and delivered without complications of PH. In future cases of noncirrhotic portal vein thrombosis in patients hoping to conceive, PVR-TIPS should be considered for definitive treatment of PH.

Keywords: Cavernous Transformation; Case Report; Non-cirrhotic; TIPS; PVT; NCPVT; Pregnancy; Hypertension

Introduction

Noncirrhotic portal vein thrombosis (NCPVT) is a rare disease affecting 3.8 of 100,000 people, with 25% being women of childbearing age.¹ It is categorized into chronic and acute forms. In chronic NCPVT, the portal vein occludes and induces cavernous transformation with collateral vein formation; this is associated with high mortality from variceal bleeding complications.² While portal vein thrombosis (PVT) more commonly occurs in patients with cirrhosis, it also occurs in noncirrhotic settings due to hypercoagulability, blood stasis, and cellular injury related to inherited hypercoagulability disorders, iatrogenic (eg prior gastric bypass surgery) or inflammatory (eg diverticulitis) processes which traumatize the portomesenteric veins.³ In some instances, the specific cause of NCPVT is not known. This case series focuses primarily on chronic NCPVT.

NCPVT significantly complicates conception for women. Pre-existing portal hypertension (PH) worsens as the fetus places pressure upon collateral veins, and increased blood volume during pregnancy strains variceal veins. Approximately 75% of patients with known varices bleed during pregnancy, accounting for 18%–50% of maternal deaths.⁴ Pregnancy outcomes are concerning, with alarming rates

of both maternal and perinatal deaths.^{5,6} Due to the high risks associated with pre-existing PH, reducing portal pressure is crucial before pregnancy.

Portal vein recanalization transjugular intrahepatic portosystemic shunt (PVR-TIPS) is a novel procedure which combines portal vein (PV) recanalization and angioplasty with transjugular intrahepatic portosystemic shunt (TIPS) to open the main PV, with the TIPS stent serving to maintain brisk outflow for mesenteric drainage.⁷ By restoring flow through the portomesenteric venous system, PVR-TIPS reduces portal pressures. The following cases involve patients with chronic NCPVT who successfully safely conceived and delivered following portal pressure reduction via PVR-TIPS.

Case Report

The first patient was a 32-year-old gravida 1 para 1 at the time of PVR-TIPS. After caesarean section for a prior pregnancy, she experienced persistent abdominal pain. Further investigation with US revealed NCPVT. Despite systemic anticoagulation, subsequent computed tomography (CT) confirmed NCPVT with cavernous transformation (Figure 1). Subsequent esophagogastroduodenoscopy (EGD) demonstrated esophageal varices. A comprehensive clinical and laboratory thrombophilia workup was unrevealing. Despite persistent anticoagulation, the patient continued to have NCPVT, and she was referred to Northwestern for PVR-TIPS.

The patient underwent PVR-TIPS without complication; hepatic and portal venous access was achieved via the right internal jugular and splenic veins (Figure 2). The patient's hepatic venous pressure gradient was decreased from 14 to 5 mmHg. Her shorter-term postprocedure course was uneventful. Her TIPS and recanalized portomesenteric system remain patent through 36-month follow-up (Figure 3). At 36-month follow-up, the patient remains anticoagulated,

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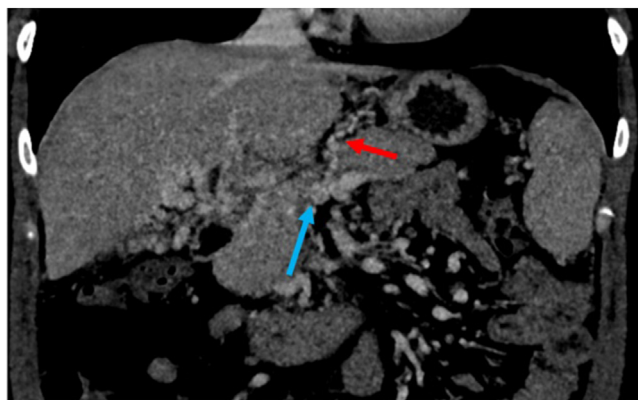


Figure 1. Pre-PVR-TIPS coronal CT demonstrates occlusion and cavernous transformation of the main portal vein (blue arrow), with upper abdominal collaterals including a prominent gastric varix (red arrow).

without interim medium- or long-term adverse events including hepatic encephalopathy. Critically, she achieved successful pregnancy following PVR-TIPS, delivering via caesarean section approximately 11 months postprocedure. While her immediate postdelivery course was complicated by retained products of conception requiring dilation and curettage complicated by persistent subacute uterine bleeding requiring anticoagulant titration, she suffered no complications of PH during or after pregnancy.

The second patient was a 38-year-old gravida 1 para 0 at the time of PVR-TIPS. The patient initially presented with significant subacute abdominal pain. Subsequent imaging revealed chronic NCPVT with cavernoma and thrombus involvement of the main PV and superior mesenteric vein

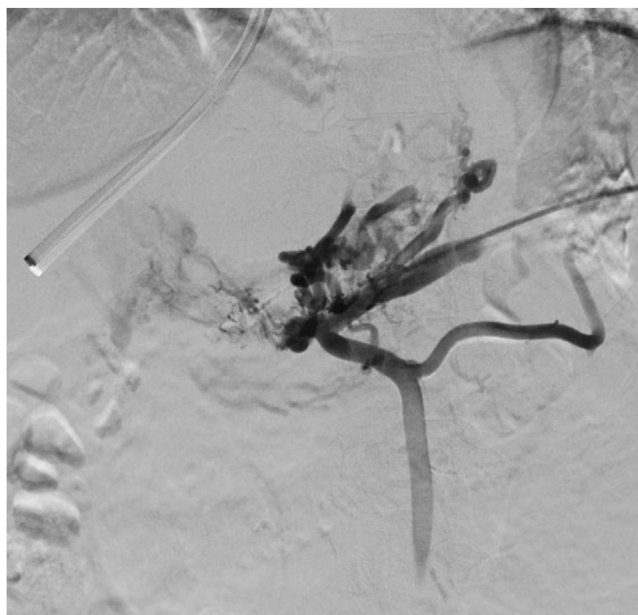


Figure 2. Initial splenic venogram via direct splenic venous access before PVR and TIPS placement confirms the CT findings of main portal vein occlusion with cavernoma.



Figure 3. TIPS venogram via internal jugular venous access 1 month after PVR-TIPS procedure confirms patency of the TIPS and recanalized main portal and splenic veins.

(SMV). EGD demonstrated esophageal varices. Laboratory workup ultimately revealed a janus kinase 2 mutation as the cause of her hypercoagulability and NCPVT. Her subsequent course included esophageal varix ligation and unsuccessful trans-splenic PVR-TIPS at an outside hospital, where they were unable to successfully cross the occluded main PV. Subsequent CT revealed persistent chronic PVT and SMV thrombosis. The patient was referred to Northwestern to reattempt PVR-TIPS.

After initial intraprocedural venograms confirmed the above CT findings (Figure 4), repeat PVR-TIPS was performed successfully via right internal jugular and direct SMV puncture (Figure 5). Her shorter-term postprocedure course was complicated by minor and intracranial hemorrhage due to overanticoagulation by her primary team. Her TIPS and recanalized portomesenteric system remain patent through 44-month follow-up (Figure 6). Subsequent EGD confirmed resolution of esophageal varices. At 44-month follow-up, the patient remains anticoagulated, without interim medium- or long-term adverse events including hepatic encephalopathy. The patient was able to achieve successful pregnancy following PVR-TIPS, delivering vaginally 26 months postprocedure without complications of PH during or after pregnancy.

Discussion

With limited literature on treatment for NCPVT in patients hoping to conceive, pregnancy is considered prohibitively risky. Current standard-of-care includes

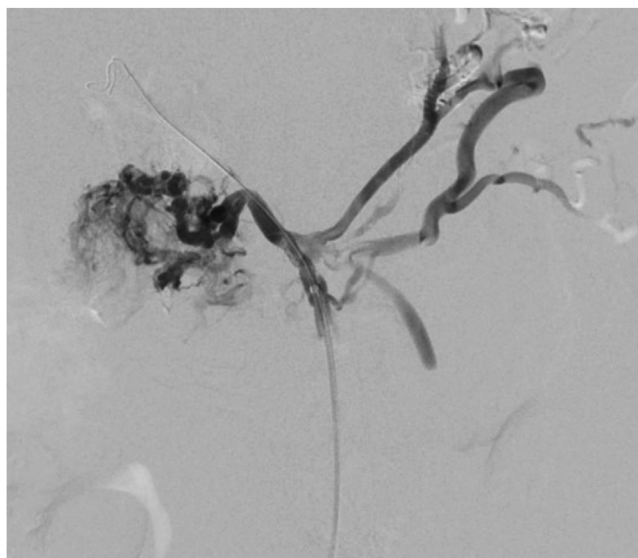


Figure 4. Initial SMV venogram via direct SMV venous access before PVR and TIPS placement confirms the preprocedural imaging findings of main portal vein occlusion with cavernoma. PVR, portal vein recanalization.

anticoagulation and beta blockers, which are not only suboptimal in treating NCPVT, but can also harm the fetus.⁸ In a prospective study evaluating anticoagulation in acute NCPVT, only 38% of patients achieved PV recanalization, with 40% developing chronic NCPVT after 1 year of anticoagulation. Because our 2 patients had progressed from acute to chronic NCPVT, pharmacologic therapy would likely prove ineffective.⁶

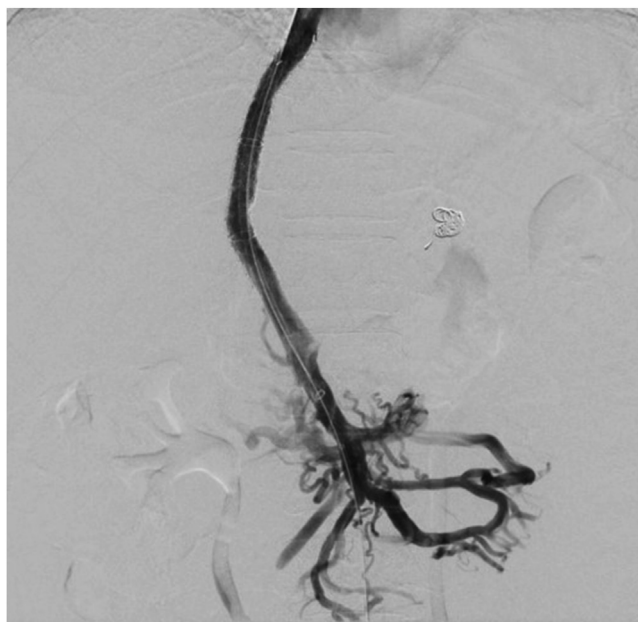


Figure 5. Venogram immediately after PVR and TIPS placement demonstrates widely patent outflow through the SMV, portal vein, and TIPS.



Figure 6. TIPS venogram via internal jugular venous access 6 months after PVR-TIPS procedure confirms patency of the TIPS and recanalized main portal vein and SMV. PVR, portal vein recanalization.

Surgical techniques, at times, may be effective. They involve either restoring native flow or providing nonphysiologic flow. The latter, less preferred due to higher rethrombosis rates, is reserved for severe cases where physiologic procedures are not infeasible.⁷ Noncirrhotic patients with underlying thrombophilia have higher recurrent thrombosis risk, worsened by hypercoagulability of pregnancy.⁹ Though surgical approaches afford higher rates of successful portomesenteric recanalization than pharmacologic approaches, risks associated with invasive surgery are deemed unacceptable by many patients. Rethrombosis after surgical revascularization remains a major limitation.

PVR-TIPS is a safe and effective minimally invasive treatment for reestablishing PV flow, in both cirrhotic and noncirrhotic patients.¹⁰ In a study of PVR-TIPS in cirrhotic PVT, technical success was achieved in 100% via novel trans-splenic technique, with 92% of recanalized PVs and TIPS remaining patent during follow-up (mean follow-up of 16.7 months).⁷ Similarly, in a recent landmark study evaluating the novel use of PVR-TIPS in NCPVT, 100% procedural success was achieved, with improvement in manifestations of portal hypertension in 87% at 6-month follow-up, and no significant long-term adverse events. At 36 months, freedom from primary TIPS thrombosis was 63%; following secondary interventions, patency increased to 81%.¹⁰ Given these outcomes, it was deemed reasonable to treat the patients in the present case series using PVR-TIPS.⁹

Strengths of this case report include the novel approach to an uncommon and difficult to treat condition.

There is the limitation of generalizability in centers still on the learning curve for PVR-TIPS. These represent the first reported cases describing patients with NCPVT who achieved successful and safe pregnancy, via novel PVR-TIPS. While additional data are needed, PVR-TIPS appears to represent a safe, effective, and addresses an unmet need for patients with chronic NCPVT who hope to become pregnant.

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IRB approval was not required for this case report.

Reporting Guidelines:

CARE.