



## Case Report

# Simultaneous Transjugular Intrahepatic Portosystemic Shunt Revision and Embolization of an Arterioportal Fistula in a Patient with Active Variceal Bleeding from Systemic Lupus Erythematosus-induced Cirrhosis

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## ABSTRACT

Transjugular intrahepatic portosystemic shunt (TIPS) occlusion is a common occurrence that can be managed based on the nature and acuity of shunt failure. Arterioportal fistulas (APFs) are rare communications between portal venous and systemic arterial vasculature that can present with symptomatic portal hypertension. In this case, we describe the management of a patient with variceal bleeding due to TIPS dysfunction complicated by the presence of an APF.

**Keywords:** Transjugular intrahepatic portosystemic shunt, Arterioportal fistula, Embolization, Endovascular intervention, Portal hypertension

## INTRODUCTION

Occlusion of transjugular intrahepatic portosystemic shunts (TIPS) is a common issue despite improvements in the techniques for TIPS creation. Failure is usually evidenced by remanifestation of symptoms of portal hypertension.<sup>[1,2]</sup> It has been shown that covered stents have a dysfunction rate of up to 44% at 2 years.<sup>[3]</sup> Arterioportal fistulas (APFs) are rare communications between systemic arteries and the portal venous system. APFs can present with symptoms of portal hypertension due to the systemic blood pressure being transmitted to the portal system. Here, we report the management of a case of TIPS dysfunction which was complicated by the presence of an incidentally detected APF.

## CASE REPORT

A 61-year-old Hispanic female underwent uncomplicated TIPS creation due to portal hypertension in the setting of systemic lupus erythematosus (SLE)-induced cirrhosis. Approximately 1 year later, she became symptomatic with ascites and was noted to have TIPS stenosis of 67% in the proximal end, which required extension of the stent with an additional 2 cm toward the hepatic venous-inferior vena cava (IVC) junction followed by venoplasty.

Approximately 3 years later, the patient presented with bleeding esophageal and gastric varices with a drop in her hemoglobin to 7.3 mg/dL from baseline of 10 mg/dL. Ultrasonography

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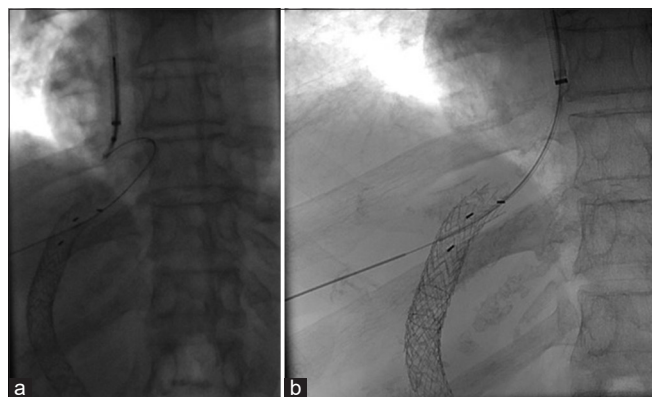
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suggested impending TIPS failure with peak TIPS velocity of 86 cm/s. A computed tomographic angiography was done, which confirmed the TIPS stenosis and revealed an incidental high flow APF between a branch of the superior mesenteric artery (SMA) and the superior mesenteric vein (SMV) causing significant dilation of the SMV.

As her portal hypertension was likely caused by two factors (TIPS occlusion and APF), we decided to treat her with TIPS recanalization along with embolization of the APF. Standard TIPS access was attempted from the right internal jugular vein, but the stent could not be accessed despite multiple attempts and techniques due to complete occlusion of the proximal end of the stent. It was then decided to access the TIPS directly through the transhepatic approach. Ultrasound and fluoroscopic guidance were utilized to access the TIPS percutaneously using a Chiba needle (Cook, Bloomington, IN). With the tip of the Chiba needle in the lumen of the stent, a 0.018" wire was able to be maneuvered through the TIPS into the right atrium. This wire was then snared from the right atrium, which helped in establishing a through-and-through access from the outside through the liver and exiting through the right internal jugular vein (Figure 1). Over this wire, a 4-Fr Cobra catheter (Cook, Bloomington, IN) was passed into the stent.

A portogram showed near-complete occlusion at the IVC-TIPS junction (Figure 2). The right atrial pressure was 4/0 mmHg (mean 2), IVC pressure was 5/5 mmHg (mean 5), and direct portal vein pressure measured 52/30 mmHg (mean 42). Serial dilatation of the TIPS stent was then performed with 5, 6, 8, and 10 mm balloons.

A post-venoplasty portogram showed interval improvement in overall stenosis but still with persistent moderate stenosis



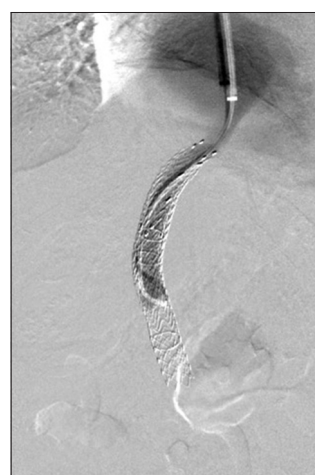
**Figure 1:** A 61-year-old female with chronic transjugular intrahepatic portosystemic shunt (TIPS) dysfunction and systemic lupus erythematosus-induced cirrhosis presented with variceal bleeding, occluded TIPS, and an arterioportal fistula. (a) 0.018" wire from the percutaneous access into the TIPS being snared from jugular access. (b) Body floss access from the outside through liver parenchyma into the TIPS and continuing through the sheath from the right internal jugular vein to the outside.

at the TIPS-IVC junction, so a 10 mm × 40 mm Fluency covered stent graft (Bard Peripheral Vascular, Tempe, AZ) was placed in the proximal portion of the TIPS and dilated with a 10 mm balloon (Figure 3). The right atrial pressure increased to 15/10 mmHg (mean 12), IVC 15/15 mmHg (mean 15), and direct portal pressure improved to 35/31 mmHg (mean 32). The portosystemic gradient was still high at 20 mmHg.

With the TIPS occlusion taken care of, the fistula between the SMA and SMV required prompt closure to bring down the portal pressure and the elevated right heart pressure



**Figure 2:** A 61-year-old female with chronic transjugular intrahepatic portosystemic shunt (TIPS) dysfunction and systemic lupus erythematosus-induced cirrhosis presented with variceal bleeding, occluded TIPS, and an arterioportal fistula. Initial injection through TIPS showing near-complete obstruction at the proximal end.



**Figure 3:** A 61-year-old female with chronic transjugular intrahepatic portosystemic shunt (TIPS) dysfunction and systemic lupus erythematosus-induced cirrhosis presented with variceal bleeding, occluded TIPS, and an arterioportal fistula. TIPS venogram following stent extension shows brisk flow.

arising from the shunting of the arterial blood into the portal system. Arterial access was obtained through the right common femoral artery. An SMA angiogram showed a high flow arterioportal shunting from the ileocolic branch of the SMA to a large varix in the SMV (Figure 4).

Selective catheterization of this branch was then performed and further oblique images were obtained. The fistula was then closed with a combination of a 9 mm microvascular plug (Medtronic, Dublin, Ireland), five 7 mm × 30 mm Concerto detachable coils (Medtronic, Dublin, Ireland), and 1 cc of Gelfoam slurry (Pfizer, New York, NY) using a sandwich technique (Figure 5).

The portal venous pressure was once more measured through the catheter in the TIPS shunt and showed significant improvement at 27/22 (mean 24). The portosystemic gradient at this time was 12 mmHg. There were no immediate complications with the procedure. At 2-month follow-up, the TIPS was accessed and showed wide patency with a TIPS pressure of 7/1 mmHg (mean 5). The right atrial pressure was 2/-6 mmHg (mean -1). The patient was seen in IR clinic 1 month later and reported no symptoms of portal hypertension, including no further episodes of bleeding.

## DISCUSSION

Despite technical improvements in TIPS creation, shunts are often subject to dysfunction at an estimated rate of 44% at 2 years.<sup>[3]</sup> Guidelines provided by the American Association for the Study of Liver Diseases state that recurrence of pH symptoms is the best indicator of shunt dysfunction.<sup>[1]</sup> Close follow-up with ultrasound is effective in the monitoring

of TIPS dysfunction as evidenced by 20–50% decrease in conduit flow, TIPS velocity <50 cm/s or >250 cm/s, or a reversal of flow direction in the intrahepatic portal branches.<sup>[4,5]</sup> Angiographic evidence of TIPS failure includes overt occlusion, portosystemic pressure gradient greater than 12–15 mmHg, or a greater than 50% reduction in diameter of the shunt.<sup>[6]</sup> Depending on the timing, TIPS failure may be bile or non-bile related, may be due to stent movement (migration, shortening, and displacement), resulting in pseudointimal hyperplasia, all of which may result in thrombosis and/or stenosis ultimately leading to occlusion and shunt failure.<sup>[2]</sup>

In this case, a 61-year-old female with long-standing liver disease had required multiple TIPS revisions and close follow-up with non-invasive imaging. Her case highlights a common feature of TIPS failure in that the location of her occlusion was predominately at the proximal end near the hepatic vein-IVC junction. Pereira *et al.* examined TIPS dysfunction in detail and proposed a helpful clinical practice algorithm to determine management course, including thorough descriptions of treatment options.<sup>[2]</sup> Stratification begins with determining the nature of failure (stenosis vs. occlusion) followed by anatomic considerations (proximal, mid, and distal) and ultimately provides suggestions for treatment based on the underlying issue. Treatment options encompass a wide range of interventional techniques and can range from angioplasty and stent revision to new shunt creation. Rescue therapy may be required in recalcitrant cases with splenic artery embolization or balloon transvenous obliteration techniques (balloon-occluded retrograde



**Figure 4:** A 61-year-old female with chronic transjugular intrahepatic portosystemic shunt (TIPS) dysfunction and systemic lupus erythematosus-induced cirrhosis presented with variceal bleeding, occluded TIPS, and an arterioportal fistula. Selective angiography of the superior mesenteric artery shows a large arterioportal fistula with the superior mesenteric vein.



**Figure 5:** A 61-year-old female with chronic transjugular intrahepatic portosystemic shunt (TIPS) dysfunction and systemic lupus erythematosus-induced cirrhosis presented with variceal bleeding, occluded TIPS, and an arterioportal fistula. Successful embolization of the arterioportal fistula with 9 mm microvascular plug, five 7 mm × 30 mm Concerto detachable coils, and 1 cc Gelfoam slurry using a sandwich technique.

transvenous obliteration/balloon-occluded antegrade transvenous obliteration).

Beyond technical considerations, in a patient such as this one with recurrent TIPS occlusion, it is important to examine the nature of the recurrence. Liver disease is extremely common in patients with SLE and may affect over half of patients suffering from this disease, the extent of which is typically mild and can be due to the inflammatory nature of SLE itself, drug-induced liver injury, fatty liver, alcohol, viral hepatitis, or autoimmune hepatitis (as in our patient).<sup>[7]</sup> In addition, there have been cases reported of non-cirrhotic portal hypertension in patients with SLE.<sup>[8]</sup> Lupus anticoagulant has been frequently associated with both arterial and venous thrombosis formation, including reports of portal vein thrombosis. This could have contributed to the recurrent TIPS occlusions in our patient. The patient was diagnosed with SLE in 2009 and has suffered from many sequelae of the disease, including pericardial effusion, pleural effusion, and oral ulcers.

APFs are rare communications between portal venous and systemic arterial vasculature. They can be congenital, traumatic, iatrogenic, malignant, infectious, or inflammatory in origin.<sup>[9]</sup> They can present with portal hypertension manifesting as variceal bleeding or recalcitrant ascites or rarely with congestive heart failure. In this case, an APF developed insidiously for 3 years between TIPS revisions such that the discovery was made on a computerized tomography scan during an admission for symptomatic TIPS failure. As SLE is strongly associated with inflammatory changes and vasculitis (including mesenteric vasculitis), we consider this disease as a possible contributor to the APF formation in this patient.<sup>[10]</sup> The APF, in this case, worsened the complications of portal hypertension from TIPS occlusion, as the fistula transmitted the systemic arterial pressure directly into the portal system.

## CONCLUSION

To resolve the symptoms in this patient, it was necessary to fix both the TIPS occlusion and the APF. The case was also made more complex by the inability to access the TIPS from jugular approach, which necessitated direct percutaneous access of the TIPS and a body floss wire technique for access into the TIPS. Our report demonstrates a successful management strategy in this unique scenario.

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Department of Interventional Radiology, Jackson Memorial Hospital.

## Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms.

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Nil.

## Conflicts of interest

There are no conflicts of interest.

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