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CASE REPORT

Transjugular Intrahepatic Portosystemic Shunt Creation in Isolated Persistent Left Superior Vena Cava and Portal Vein Thrombosis

Sara Rostami, Ryan Dunn, Derek Rubadeux and Ali Kord

Division of Interventional Radiology, Department of Radiology, University of Cincinnati, USA

Abstract:
Isolated persistent left superior vena cava is a rare congenital venous anomaly. It imposes technical challenges and increased risks in patients requiring a transjugular intrahepatic portosystemic shunt. The patient was a 67-year-old man with cirrhosis, recurrent large-volume ascites, hepatic hydrothorax, and portal vein thrombosis. The patient had a history of failed transjugular intrahepatic portosystemic shunt creation using a conventional CO_2 portal venography technique via a left jugular vein access. The patient underwent successful transjugular intrahepatic portosystemic shunt creation under fluoroscopy and intravascular ultrasound guidance. The patient required transjugular intrahepatic portosystemic shunt revision with mechanical thrombectomy in 2 months. Intravascular ultrasound can provide additional live information to assist transjugular intrahepatic portosystemic shunt creation in patients with complex congenital venous anatomy, including those with isolated persistent left superior vena cava.

Keywords:
persistent left superior vena cava, transjugular intrahepatic portosystemic shunt, intravascular ultrasound, portal hypertension

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Introduction

Persistent left superior vena cava (PLSVC) is present in 0.3%-0.5% of the general population, where the left-sided superior vena cava persists beyond the final stages of fetal development [1]. Most cases with PLSVC (90%) have double SVC, where the anomalous left SVC is accompanied by a right SVC that either runs independently along the mediastinum or joins the left SVC before emptying into the coronary sinus [2]. In the remaining 10% of PLSVC cases, the right SVC is absent and referred to as an isolated PLSVC [1, 2].

Transjugular intrahepatic portosystemic shunt (TIPS) creation is often performed via a right internal jugular (IJ) vein access. In patients with an isolated PLSVC, the TIPS creation should be performed from a left IJ vein approach and through the coronary sinus, which may increase the risk of cardiac arrhythmia [3]. In addition, the angulation through a left IJ venous access may limit certain maneuvers to obtain and maintain access to the portal vein during the TIPS creation [3]. The thrombosed portal vein in patients with an isolated PLSVC may impose more challenges because the conventional TIPS using CO_2 portal venography may not opacify the thrombosed portal vein branches [4]. Intravascular ultrasound (IVUS) provides live visualization of the venous structures and helps with needle direction to access the portal vein during the TIPS creation [5]. In this manuscript, we describe a case of isolated PLSVC with portal vein thrombosis undergoing an IVUS-guided TIPS creation for refractory ascites and hepatic hydrothorax.

Case Report

The Institutional Review Board exempted this case report from the informed consent requirement for publication. The patient was a 67-year-old man with alcoholic cirrhosis complicated by recurrent large-volume ascites and right hepatic hydrothorax, non-bleeding esophageal varices, and nonocclusive portal vein thrombosis on apixaban. The patient declined a liver transplant evaluation. The Model for End-Stage Liver Disease score was 16, with a Child-Pugh score of class B. The patient's preoperative contrast-enhanced

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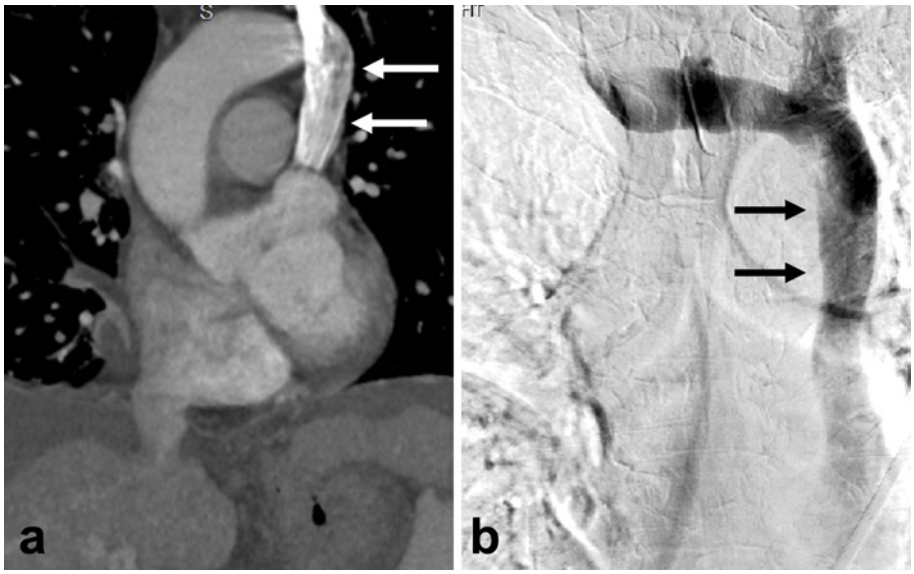

Figure 1. Contrast-enhanced CT (a) and venography (b) show an isolated PLSVC draining into the coronary sinus. There is no right SVC.

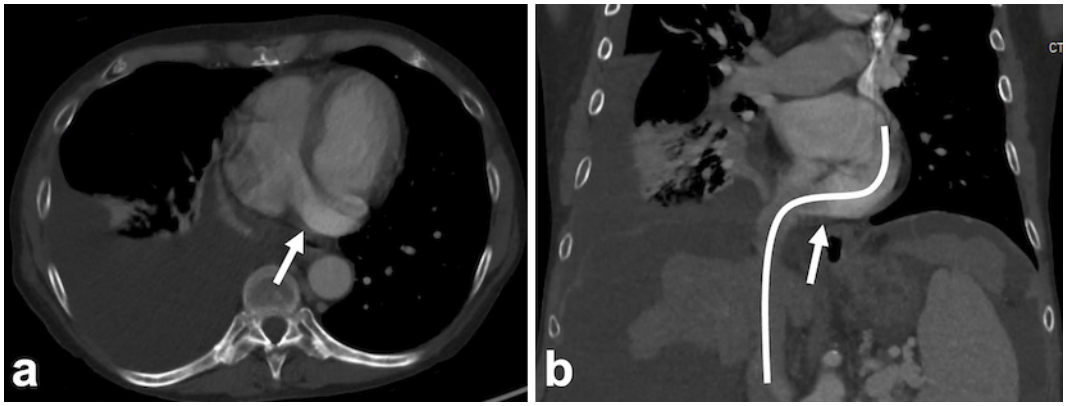

Figure 2. Axial (a) and coronal (b) contrast CT of the chest illustrating the extra curve of the sheath (white curved line, b) while passing the coronary sinus (white arrows, a and b).

computed tomography (CT) of the abdomen showed hepatic cirrhosis with large right pleural effusion and ascites and portal vein thrombus within the main and right portal vein. The patient had a history of unsuccessful TIPS creation using the conventional technique, which involved CO_2 portal venography via left jugular vein access (**Fig. 1**). The initial right IJ demonstrated an isolated PLSVC. Therefore, a left IJ access was obtained. A cardiac MRI was performed after the procedure to assess the cardiac and great vessel anatomy better, confirming an isolated PLSVC draining into the coronary sinus. The patient was referred to the authors' institution for a TIPS placement under IVUS guidance.

The patient underwent a TIPS procedure under general anesthesia. Given the known isolated PLSVC, left IJ vein access was obtained under US guidance and dilated to accept a 10 Fr Sheath (Cook Medical, Bloomington, IN) for TIPS placement. The anesthesia team was notified while passing, and wires, catheters, and sheaths were passed through the heart to monitor for arrhythmia. No cardiac arrhythmia was observed while passing the wires and sheath through the heart in this case (**Fig. 2**). Similarly, a left common femoral vein access was obtained under US guidance and was dilated to accept a 9-Fr sheath (Cordis, Santa Clara, CA) to be utilized for intravascular ultrasound. An 8-F intravascular probe probe (AcuNav, Siemens, Malvern, PA) was advanced to the inferior vena cava (IVC) and used to identify the hepatic and portal veins. There was a large clot burden within the main portal vein and the right portal vein branches.

Based on the live IVUS evaluation and the preprocedural CT images, a middle hepatic vein to right portal vein access was thought to provide a smoother path and, therefore, was selected. Through the IJ access sheath, a 5-Fr multipurpose catheter (MPA, Angiodynamics, Latham, NY, USA) was used to select the middle hepatic vein under IVUS and fluoroscopic guidance, and the 10-Fr sheath was advanced into the middle hepatic vein. Access into the right portal vein was obtained using an access needle (Colapinto, Cook Medical, Bloomington, IN, USA) through the middle hepatic vein under fluoroscopic and intravascular ultrasound

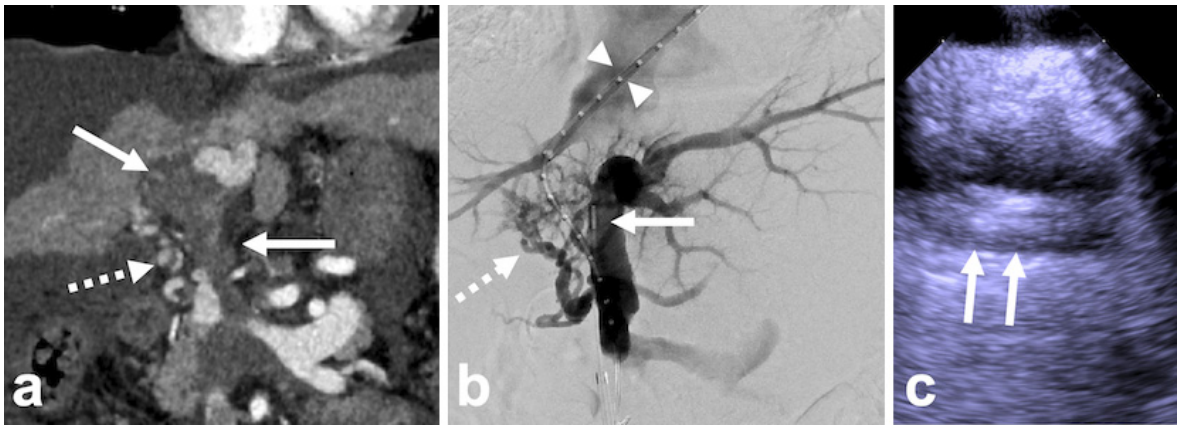

Figure 3. Contrast-enhanced CT before the TIPS procedure (a) shows complete thrombosis of the main and right portal vein (solid arrows) with multiple collaterals (dotted arrow). Dual portal and hepatic venography (b) demonstrating similar findings with multiple collaterals (dotted arrow). The extra curve of the sheath (arrowheads) is caused by an isolated LPSVC, which helps with hepatic vein access. IVUS (solid arrow, b) is placed in the IVC via groin access and shows large clots (solid arrows, c) within the portal vein.

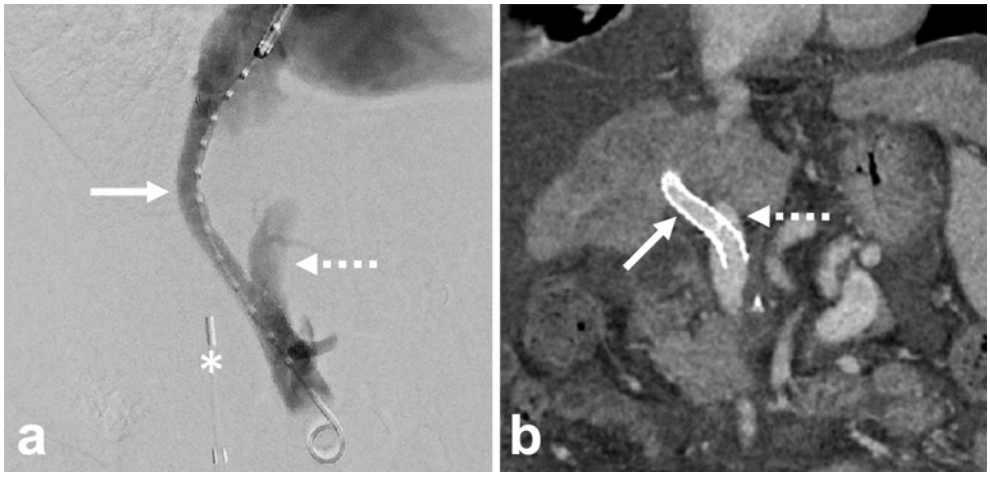

Figure 4. The portal venography (a) and contrast-enhanced CT (b) after TIPS creation show a patent TIPS stent (solid arrows) and a left portal vein (dotted arrows). Asterix marks the IVUS probe within the inferior vena cava.

guidance. A 5 Fr hydrophilic catheter (Glidecath, Terumo, Somerset, NJ, UK) and 0.035" stiff guide wire (GLIDEWIRE, Terumo) were used to get across the portal vein thrombosis. Portal venography demonstrated similar findings of portal vein thrombosis with multiple collaterals (**Fig. 3**). It was difficult to pass the 10 Fr sheath to the portal vein branch using a 0.035" stiff guide wire (Amplatz, Boston Scientific, Marlborough, MS, USA). Therefore, a 0.035" extra-stiff guide wire (Lunderquist, Cook Medical) was used to straighten the path and to decrease the angulation which was helpful to advance the sheath into the portal vein. An 8-10 mm × 10 cm (8 cm covered and 2 cm uncovered) controlled expansion stent graft (Viatorr, Gore, Flagstaff, AZ, USA) was deployed under fluoroscopic and IVUS guidance (**Fig. 4**). The stent graft was post-dilated to 8 mm using a balloon (Conquest, BD, Franklin Lakes, NJ, USA). A portal venogram demonstrated a patent portal vein and TIPS stent with a brisk flow (**Fig. 3**). The portosystemic gradient decreased from 16 to 3 mmHg. All the catheters and wires were removed.

The patient was observed overnight and was discharged on apixaban 2.5 mg twice daily (Pfizer, New York, NY, USA) the day after the procedure. The patient's condition initially improved after the TIPS creation with decreased hepatic hydrothorax but worsened at 1 month. A Doppler US exam of the TIPS was performed one month after the procedure as part of a follow-up clinic visit, which showed a patent TIPS stent but with increased velocities up to 254 cm/s, suggestive of narrowing. The patient did not undergo a TIPS evaluation at that time because he had myocardial infarction due to coronary artery disease 3 weeks after the procedure and was instructed to postpone any non-urgent procedure. The patient returned to the emergency room two months after the TIPS procedure with shortness of breath and abdominal distension and was found to have thrombosis of the splenic and superior mesenteric veins to the intrahepatic por-

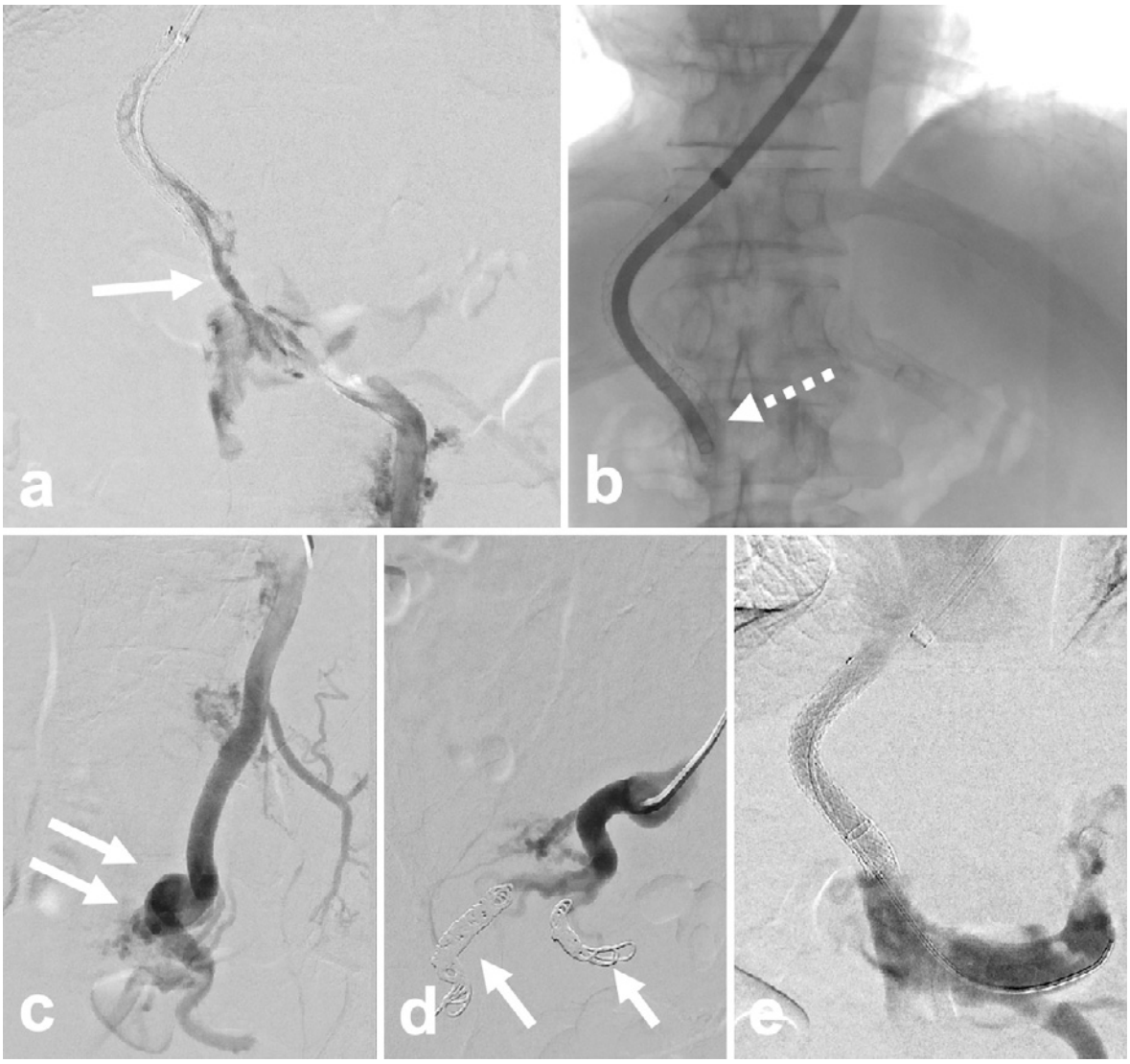

Figure 5. Follow-up portal venography (a) shows a thrombosed portal vein and TIPS stent (solid arrow). Mechanical thrombectomy was performed (dotted arrow, b). A large portosystemic shunt (solid arrows, c) is visualized, which is embolized with coils (solid arrows, d). Final portal venography shows patent portal vein and TIPS stent (e).

tal veins and occluding the TIPS stent.

The patient underwent a TIPS revision with mechanical thrombectomy of the splenic, superior mesenteric, and main portal veins and the TIPS stent (**Fig. 5**). The procedure was performed under general anesthesia. A 16 Fr Sheath (Cook Medical, Bloomington, IN, USA) was placed into the left IJ vein under ultrasound and fluoroscopic guidance. The TIPS stent was selected using a curved catheter (MPA, Angiodynamics, Latham, NY, USA). Initial venogram showed extensive thrombosis within the splenic, superior mesenteric veins, portal veins branches, and the TIPS stent (**Fig. 5**). Mechanical aspiration thrombectomy was performed using a thrombectomy catheter (Cat-16 Flash lightening, Penumbra, Alameda, CA, USA). A hepatofugal flow was present in the inferior mesenteric vein with a large portosystemic shunt. The shunt was subselected using a 5 F catheter and was embolized using coils to help with the portal vein flow (**Fig. 5**). The final venogram showed patent splenic and portal veins with improved flow through the TIPS stent. The patient was admitted to the hospital and was started on IV anticoagulation after the procedure. US exam of the TIPS after the procedure showed a patent TIPS stent and portal vein.

The patient's symptoms, including ascites and hepatic hydrothorax, slightly improved after the TIPS revision. The patient's clinical course was later complicated by hyponatremia, and he expired 4 months after the initial TIPS procedure.

Discussion

TIPS creation in patients with an isolated PLSVC using fluoroscopy with or without IVUS guidance has been described before [3, 6]. Portal vein thrombosis and an isolated PLSCV made the patient in this study more challenging compared to the previously reported cases [3, 6]. The TIPS procedure was successfully performed under IVUS guidance. In recent portal vein thrombosis cases, the thrombosed portal vein can be localized by IVUS and targeted by the access needle. The complementary live visualization of the venous structure by IVUS is particularly helpful in cases

with an isolated PLSVC [3]. In addition, IVUS guidance during TIPS creation can potentially decrease the procedure time, radiation dose, and needle passes [7]. IVUS can provide live visualization of portal vein branches in patients with acute or subacute thrombosis. In patients with chronic portal vein thrombosis, the portal vein may be diminutive with numerous collaterals, and the IVUS may have limited utility in portal vein access.

The isolated PLSVC in our patient was draining into the coronary sinus and right atrium, providing a path to advance the sheath to the IVC and hepatic veins. Isolated PLSVC may be associated with an unroofed coronary sinus or atrial septal defect in 50%-70% of the cases [1]. In such cases, advancing the sheath may worsen the atrial septal defect and cause a right to left shunt after TIPS creation [8]. The isolated PLSVC may sometimes be drained into the left atrium, conferring a transjugular approach for portosystemic shunt creation. The routine echocardiography performed before TIPS creation may fail to diagnose isolated PLSVC and the associated cardiac abnormalities [3]. A dedicated cardiac magnetic resonance imaging may be needed to assess the anatomy in very complex cases.

Compared to the routine right IJ approach, the TIPS creation in patients with an isolated PLSVC may be associated with a higher risk of cardiac arrhythmia. A stiffer guidewire may be required to maintain portal vein access. In patients with an isolated PLSVC, transient arrhythmia may occur while passing wires, catheters, and sheath through the dilated coronary sinus, which requires extra attention by the operator and the anesthesiologist. Close communication between the operator and the anesthesia team is critical when passing wires, catheters, and sheaths through the heart. Hydrophilic curved catheters and wires help direct and pass the heart. If there is any resistance, the catheter location should be checked in contrast rather than forcefully pushing. Compared to the usual right IJ approach, the extra curve of the catheters from the left IJ approach provides better angulation to access the hepatic veins [3]. However, it provides less support for the craniocaudal pushability of the access needle and the sheath. An extra stiff guidewire was advanced in the portal vein after obtaining the portal vein in the patient in this study, which helped advance the sheath to the portal vein in preparation for stent placement. When CO_2 portal venography cannot be performed, and IVUS guidance is unavailable, a 3D roadmap using a contrast-enhanced CT [9, 10] or coexisting arterial guidance [11, 12] may be alternatives. Compared to IVUS, these methods may increase the radiation dose and procedure time. When a transjugular approach is not available or fails, other techniques may be considered to create a portosystemic shunt in patients with congenital venous anomaly such as a transfemoral, transmesenteric, transhepatic, trans-splenic, or direct transcaval approach [13, 14].

The patient in this study returned with portal vein and TIPS stent thrombosis requiring thrombectomy and TIPS revision. The revision rate after TIPS creation is often higher in patients with an isolated LPSVC than those with normal right SVC [3], though the underlying cause is unclear.

In summary, a case of successful TIPS creation in a patient with a rare anomaly of isolated PLSVC is reported. IVUS can be used as a complementary tool to assist TIPS creation in patients with complex congenital venous anatomy, particularly those with isolated PLSVC and recent portal vein thrombosis.

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Author Contribution: AK designed the study; SR, RD, DR, and AK wrote, revised, and reviewed the manuscript; SR and AK completed data analysis and image interpretation; SR, RD, and DR performed the literature search and drafted the manuscript.

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