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CASE REPORT

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Transhepatic approach of patent foramen ovale closure in the setting of congenital inferior vena cava anomaly

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Key Clinical Message

Patent foramen ovale (PFO) is the most common interatrial septal abnormality. The indications for PFO device closure are still being evaluated, with the most common reason being to prevent cerebrovascular accidents (CVA) caused by paradoxical embolism of deep vein thrombosis (DVT) in the lower extremities. This procedure is usually performed through percutaneous intervention using femoral vein access. Here, we present a case of PFO closure using a transhepatic approach, as femoral vein access was not feasible due to an interrupted inferior vena cava (IVC). The patient had a prominent left-sided IVC, larger than the right-sided IVC, and the left-sided IVC served as the main draining conduit via the hemiazygous system, which then connected to the azygous vein and emptied into the right atrium (RA). Cardiac MRI confirmed these findings, including the continuation of the suprahepatic IVC to the right atrium. With the assistance of interventional radiologist, transhepatic access was achieved, and the PFO was successfully closed. Hemostasis was achieved using coil embolization, and there were no post-procedural complications.

KEYWORDS

congenital heart disease, IVC anomaly, patent foramen ovale, stroke, vascular anomaly

1 | INTRODUCTION

Embryological development of the anomalous inferior vena cava (IVC) is a rare congenital abnormality that affects approximately 4%.[1] These malformations can present as isolated conditions or can be associated with other congenital cardiac anomalies (0.6%–2%).[2] The indications for PFO device closure are still evolving, with the most common reason being to prevent CVA caused by paradoxical embolization of DVT in the lower extremities.[3] This procedure involves percutaneous intervention using femoral vein access. However, in patients with chronic DVT, congenital anomalies of the IVC, or occlusion of the femoral vein for any reason, femoral vein access may not be possible. Here, we present a case of PFO closure using a transhepatic approach due to a congenital anomaly of the IVC.

2 | CASE PRESENTATION

A 60-year-old male presented with an acute onset of left-sided weakness and dysarthria. His past medical history includes Type 2 diabetes mellitus, hypertension, illicit

drug abuse (cocaine), and rheumatoid arthritis. The patient is noncompliant with medications. Vital signs including blood pressure, heart rate, respiratory rate, and saturation were normal. On physical examination left hand grip was weak and had mild dysarthria, otherwise was unremarkable. HbA1c is elevated at 8.9%, LDL at 79 mg/dL, and the urine drug screen is positive for cocaine. A CT head without contrast did not show acute disease, but a CTA head suggested a possible left distal PCA occlusion. CTA neck revealed bilateral carotid plaques at the bifurcation, each <50%, but appeared soft. MRI brain revealed scattered embolic-appearing infarcts affecting the right frontal lobe and sluggish flow in the distal right MCA branches. Echocardiogram showed preserved left ventricular ejection fraction (LVEF) at 73% without obvious regional wall motion abnormalities. The agitated saline bubble study is positive for a right-to-left intra-atrial shunt. Lower extremity venous Doppler ruled out DVT. The patient was discharged with a 30-day heart monitor, which came back negative for atrial fibrillation/flutter. Hyper-coagulable workup is negative. Transesophageal echocardiogram (TEE) showed an unremarkable left atrial appendage, positive agitated saline bubble study, and a PFO with bidirectional shunting across the atrial septum (see TEE Figure 1).

The RoPE (Risk of Paradoxical Embolism) score is 6, suggesting a 60% chance that the stroke was due to PFO, with an additional 8% risk of recurrent stroke/TIA within 2 years. The patient is scheduled for PFO closure, but the first attempt is unsuccessful due to a congenital anomaly with a prominent left-sided inferior vena cava (IVC) that is larger than the right-sided IVC (see Figure 2 and Video S1). Multiple angulations of an inferior venacavogram are performed, revealing that the right-sided IVC is smaller than normal, while the left-sided IVC appears much larger in size and serves as the main draining conduit through the hemiazygous system into the azygous vein and then into the right atrium. Additionally, it is observed that the right-sided infrahepatic portion of the IVC connects with the hemiazygous system rather than the right atrium. Cardiac MRI (see Figure 3) confirms the aforementioned findings, including the continuation of the suprahepatic IVC into the right atrium. With the assistance of interventional radiologist, transhepatic access is obtained, and the PFO is successfully closed. Coil embolization of the transhepatic tract is performed to achieve hemostasis after the removal of the vascular sheath. The following day, focal ultrasound of the right upper abdomen does not demonstrate a subcapsular hematoma, and the patient is discharged home in stable condition. The patient had 6 months and 1 year follow-up with no long-term complications.

3 | PROCEDURE

The patient was brought to the catheterization lab and placed on the table in the usual sterile manner. The liver area was prepped and draped for transhepatic access. General anesthesia was administered. TEE was performed to identify the location and anatomy of the PFO. The activated clotting time (ACT) was maintained around 250 seconds.

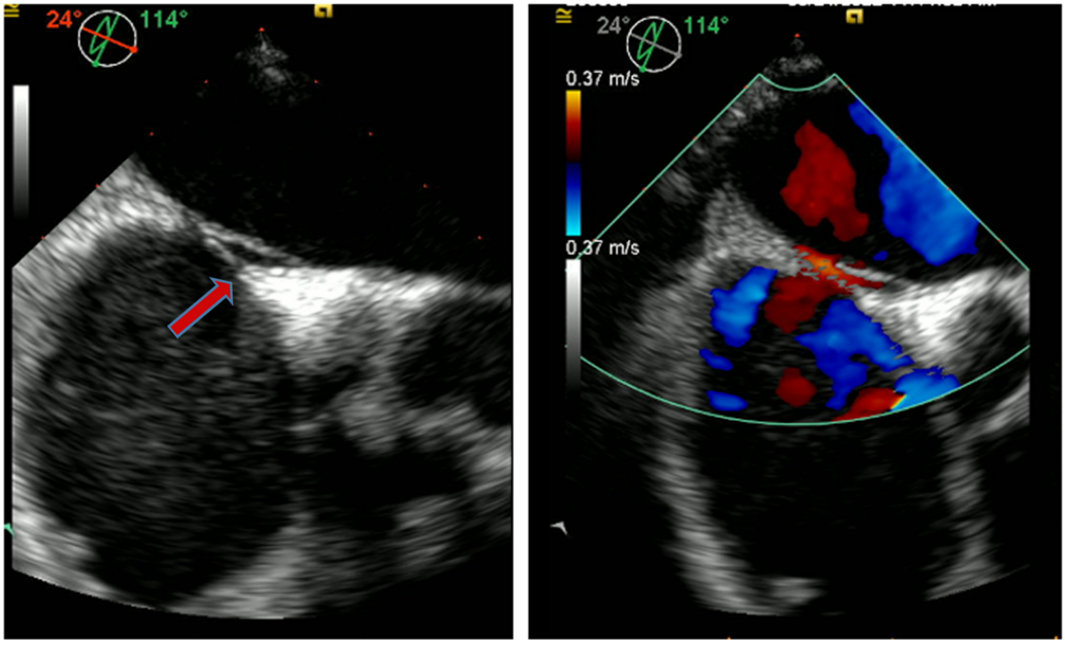

FIGURE 1 A 2D transesophageal echo image (114°) of a patent foramen ovale (PFO; red arrow) with shunting evident on the color flow Doppler.

Right hepatic vein access achieved under ultrasound guidance. A small amount of contrast was injected to obtain a venogram, which confirmed the patency of the right hepatic vein. The right hepatic vein drained into the right atrium via a short suprahepatic IVC (see Video S2). Through the multipurpose catheter, a right superior pulmonary vein angiogram was performed, demonstrating drainage into the left atrium. Based on the anatomical interrogation and with the terminal in situ, we proceeded with the deployment of a 30-mm Cardioform device, which was prepared in the routine fashion. The device was advanced over the monorail system up to the left atrium. Under the guidance of TEE and fluoroscopy, the Cardioform 30 mm device was gradually deployed (see Video S3). Subsequently, the device was "locked" in a routine fashion as well. The patient had received loading doses of aspirin and clopidogrel.

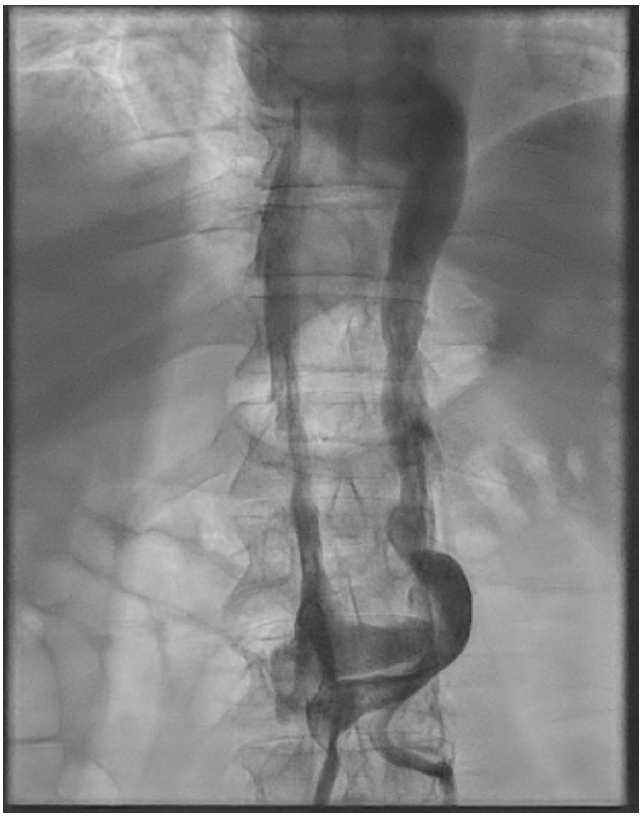

FIGURE 2 Inferior venacavogram showing dual IVC.

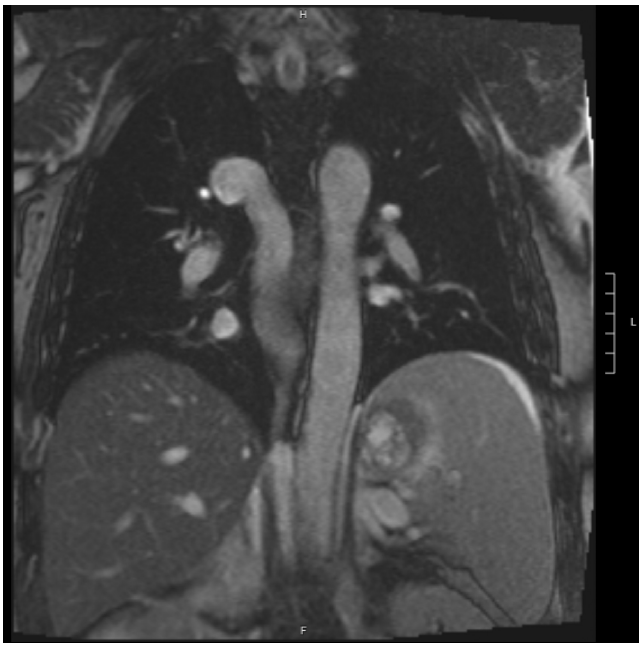

FIGURE 3 Cardiac MRI confirms the interrupted IVC.

To achieve hemostasis, embolization coils were deployed in the peripheral aspect of the right hepatic vein and in the transhepatic tract (see Figure 4). A repeat focal ultrasound of the right hepatic lobe did not reveal a subcapsular hematoma. A sterile dressing was applied to the puncture site in the right abdomen. The patient tolerated the procedure well.

4 | DISCUSSION

In younger individuals, 35% of ischemic strokes have a cryptogenic etiology. A study[4] included 60 adult patients under 55 years of age with an ischemic stroke and a normal cardiac examination. These patients were compared to a control group consisting of 100 patients.

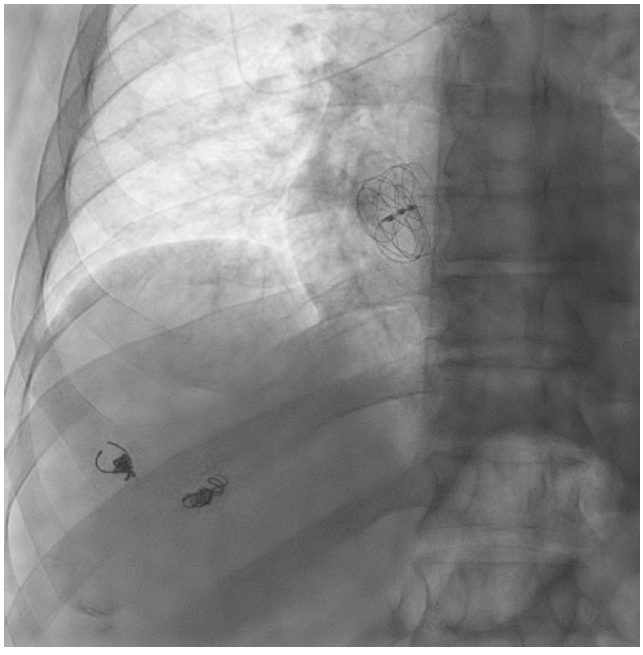

FIGURE 4 Showing coil embolization in the transhepatic tract and Cardioform device.

The prevalence of PFO was significantly higher in the stroke patients (40%) compared to the control group (10%, $p < 0.001$). The most common indication for PFO device closure is to prevent CVA caused by lower extremity DVT. The standard approach is transfemoral access. However, in conditions where femoral access is not feasible due to chronic lower extremity DVT involving the femoral vein, a large clot burden, or a congenital anomaly of the IVC and femoral veins, alternative access should be considered, including the internal jugular vein and transhepatic approach. Transhepatic permanent pacemaker lead placed in a patient with fibrosing mediastinitis and superior vena cava occlusion where traditional approach was not feasible.[5] Another case reported using transhepatic approach for atrial fibrillation ablation in the patient with heterotaxy syndrome associated with absence of IVC[6] and very few cases reported using transhepatic approach for various cardiac procedures including diagnostic and interventional cardiac catheterization, pacemaker placements, and electrophysiological procedures in pediatric and adult populations.

Similarly, there are very few cases reported using internal jugular approach for PFO closure, one of them had thrombosis of IVC with prominent venous collaterals and dilated hepatic vein, transhepatic approach was not considered. PFO closure through right internal jugular was technically difficult, the most challenging stages advancing catheter through PFO due to the acute angle.[7] No cases found using subclavian approach as not favorable due to the introduction of the thin-walled sheath through the junction of the clavicle and the first rib, which carries a high risk of bleeding.[8]

In terms of safety, the reported complications rate is less than 5% and potential complications that may arise include hemorrhage, gallbladder perforation, hepatic vein thrombosis, infection, and pneumothorax. Identifying proper anatomical landmarks and using imaging modalities such as ultrasound and fluoroscopy, along with appropriate patient selection criteria can help minimizing complications associated with this approach.[9] The technical aspect of this procedure is that the hepatic vein drains into the right atrium from a more posterior direction, which can cause more anterior transseptal puncture than expected. Right pulmonary vein ablation procedures through a transhepatic approach is a challenge due to the steep curve and this may not be challenging to reaching left atrium through PFO.[6]

Our patient was admitted with CVA and was found to have a PFO. Due to the high RoPE score, the patient was referred for PFO device closure to prevent recurrent CVAs. Our patient has interruption of the inferior vena cava (IVC) at the infrahepatic/suprarenal segment. Below the level of the renal veins, a right-sided IVC is observed. At the level of the renal veins, the IVC becomes interrupted, with the continuation of the IVC by prominent azygous and hemiazygous veins, which the renal veins drain into. In the suprahepatic region, the IVC is seen extending into the right atrium. The hepatic veins extend into the suprahepatic IVC, and large azygous and hemiazygous veins are observed. This is consistent with interruption of the IVC and azygous or hemiazygous continuation (see Figure 5).[10] It is crucial to understand the anatomical arrangements in percutaneous cardiac interventions. Our patient had a successful PFO closure through a percutaneous transhepatic approach. One potential complication of the transhepatic approach is bleeding at the noncompressible site.[11] Vascular coils have been used to achieve successful hemostasis. Vascular plugs are a recent addition to interventional cardiology closing devices and have been successfully used to occlude large vessels or conduits.[8]

The largest study on this approach in adult patients, comprising six patients, applied to management of cardiac arrhythmias and venous anomalies showed that the technique could be applied safely.[12]

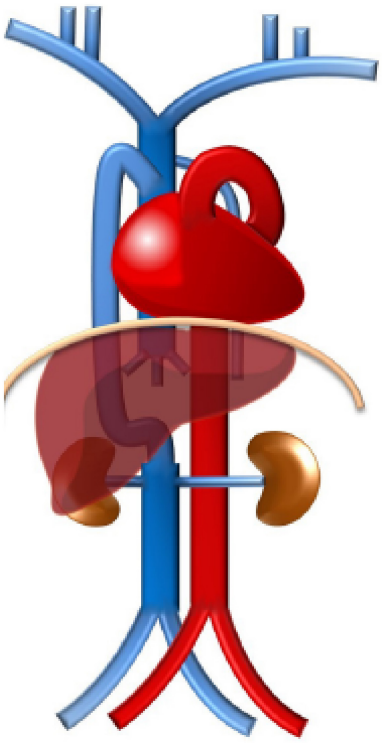

FIGURE 5 Interruption of the hepatic segment of the IVC/Azygos continuation of the IVC schematic. In this anomaly, the hepatic segment of the IVC is absent or hypoplastic, with the hepatic veins draining into the right atrium via the suprahepatic IVC.

5 | CONCLUSION

With the successful PFO closure using the transhepatic approach in our patient, along with few other case reports utilizing the same approach, we can report that this approach is less traumatic, well-tolerated, and technically less challenging compared to the jugular approach in patients where femoral access is not feasible due to congenital abnormalities of the IVC, a large clot burden in the femoral veins, and other factors. It is crucial to remember that this method should be carried out in conjunction with an experienced interventional radiologist. Identifying proper anatomical landmarks and using imaging modalities such as ultrasound and fluoroscopy, along with appropriate patient selection criteria can help minimize complications associated with this approach.

AUTHOR CONTRIBUTIONS

Deepa Soodi: Writing – review and editing. **Somto Nwaedozie:** Writing – review and editing. **Milind Shah:** Supervision. **Sudhakar Girotra:** Supervision.

FUNDING INFORMATION

None.

CONFLICT OF INTEREST STATEMENT

The authors have no conflict of interest.

DATA AVAILABILITY STATEMENT

This is a case report, there is no data or link to repository available.

CONSENT

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

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SUPPORTING INFORMATION

Additional supporting information can be found online in the Supporting Information section at the end of this article.

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