

Transhepatic device closure of atrial septal defect in children associated with interrupted inferior vena cava

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ABSTRACT

Percutaneous transcatheter closure of fossa ovalis atrial septal defect (FOASD) is an established procedure. When femoral venous approach is unfeasible due to interrupted inferior vena cava (IVC) or occluded femoral veins, other interventional methods through transjugular, transhepatic approach, or surgical closure can be performed. We report two cases of transhepatic device closure of FOASD associated with IVC interruption successfully performed without any complications.

Keywords: Atrial septal defect, interrupted inferior vena cava, septal occluder, transcatheter closure, transhepatic approach

INTRODUCTION

Atrial septal defects contribute to 8% of congenital heart defects.^[1] Both transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) play a crucial role in assessing the suitability for transcatheter closure. Inferior vena cava (IVC) interruption or bilateral femoral vein occlusion may hinder the interventional approach through the femoral route. In such situation, surgery and percutaneous closure using transjugular or transhepatic route are the options.^[2-4] Although transhepatic approach is well described but less practiced. Hereby, we report two cases of large fossa ovalis atrial septal defect (FOASD) with IVC interruption who underwent successful transcatheter device closure through the transhepatic route.

CASES

Case 1

A 9-year-old girl weighing 18 kg underwent repair of congenital diaphragmatic hernia during neonatal

period. She was diagnosed to have FOASD and referred for treatment. Examination revealed saturation of 99%, prominent parasternal pulsations, wide fixed split S2, and grade 2/6 ejection systolic murmur over left parasternal area. Chest X-ray showed cardiomegaly with increased pulmonary vascularity. Electrocardiogram showed right axis deviation and incomplete right bundle block. TTE and TEE revealed large 25-mm FOASD with small retro aortic rim. There was IVC interruption draining into the azygos vein.

Case 2

A 4-year-old asymptomatic boy weighing 16 kg was diagnosed to have FOASD. Examination revealed wide fixed S2 and 2/6 systolic murmur. TTE and TEE showed a 19-mm FOASD with left-to-right shunt. The posterior rim was floppy and measured 4 mm. The child was planned for transcatheter device closure under general anesthesia. Femoral venous access could not be obtained due to difficulty in advancing the guidewire.

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Angiogram through cannula showed IVC interruption at the infrahepatic level and draining into the azygos vein through multiple venovenous collaterals. Thus, the procedure was abandoned and rescheduled through transhepatic approach.

Procedure

The procedure was done under intubation general anesthesia after obtaining informed consent. The right thoracoabdomen was draped for the hepatic access. Coagulation profile and bleeding time were done before the procedure.

i. Hepatic vein access: An ultrasound assessment of the liver and hepatic veins was done to select a straight course of hepatic vein from the right side of the chest to IVC [Figure 1a and b]. The ultrasound transducer was covered with a sterile polythene sleeve and Tegaderm (3M science, St. Paul, MN, US). Then, transhepatic access was taken using 20 G puncture needle by Seldinger’s technique under ultrasound guidance [Figure 1c]. Apnea was induced during the access to prevent liver injury due to respiratory movements. Hand contrast injection was given to confirm the hepatic vein course. After confirming the access, a hydrophilic angled guidewire was introduced and exchanged for a 5-Fr short sheath. Intravenous unfractionated heparin 100 units/kg was given. A baseline hemodynamic assessment including Qp, Qs, and pulmonary artery pressures was measured using 5-Fr multipurpose catheter.

ii. Device closure: Case 1: After an initial assessment by TEE [Figure 2a and b], a 5-Fr multipurpose catheter was advanced through a short sheath from the hepatic access and crossed the FOASD. The catheter was positioned in the left upper pulmonary vein (LUPV) and exchanged with 0.035” Amplatzer extra stiff wire. Then, the short sheath was removed and hepatic access track was gradually dilated using 8-Fr and 10-Fr short dilators. A 10-Fr flexor sheath (Cook Medical, Bloomington, MN, US) was positioned over the wire in the LUPV and confirmed with hand injection [Figure 1d]. A 28-mm Amplatzer septal occluder (ASO) (Abbott medical MN, US) was deployed under TEE and fluoroscopy guidance [Figure 2c and d]. Case 2: Essentially, all steps were similar to the case 1 except for a 22-mm ASO was selected for the 19-mm large defect [Figure 3a]. However, the device could not be aligned to the interatrial septum and prolapsed through the defect. Then, the occluder was removed and delivery sheath was positioned in the right upper pulmonary vein (RUPV) using multipurpose catheter and guidewire assembly to have good alignment of the occluder. Then, ASO was again gradually deployed from the RUPV [Figure 3b and c]. Finally, the device was well-positioned and aligned to the interatrial septum.

iii. Coil closure of hepatic track: After the deployment of the device, a 5-Fr multipurpose catheter was advanced over the wire into the flexor sheath. The

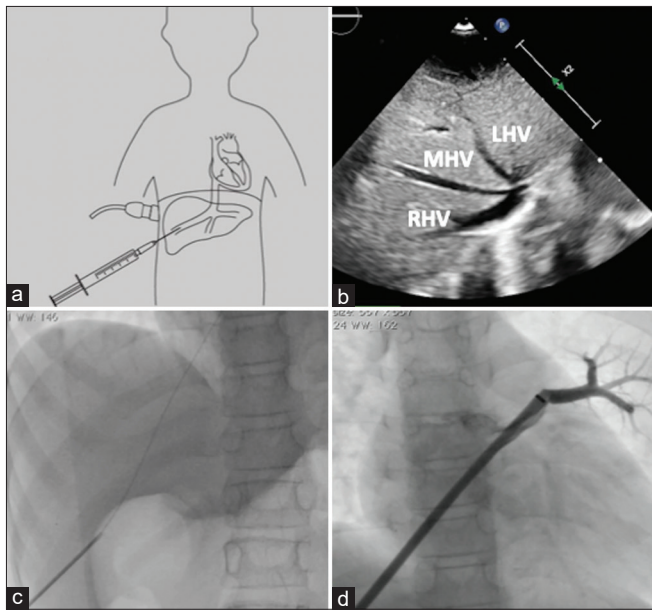


Figure 1: (a) Schematic diagram showing ultrasound-guided hepatic vein access. (b) Ultrasound abdomen image showing branches of hepatic veins. (c) Plain fluoroscopy image showing puncture needle with hydrophilic wire in the hepatic vein reaching RA. (d) Hand injection through straight flexor sheath showing left upper pulmonary vein and its branches. RA - Right atrium, LHV - Left hepatic vein, MHV - Middle hepatic vein, RHV - Right hepatic vein

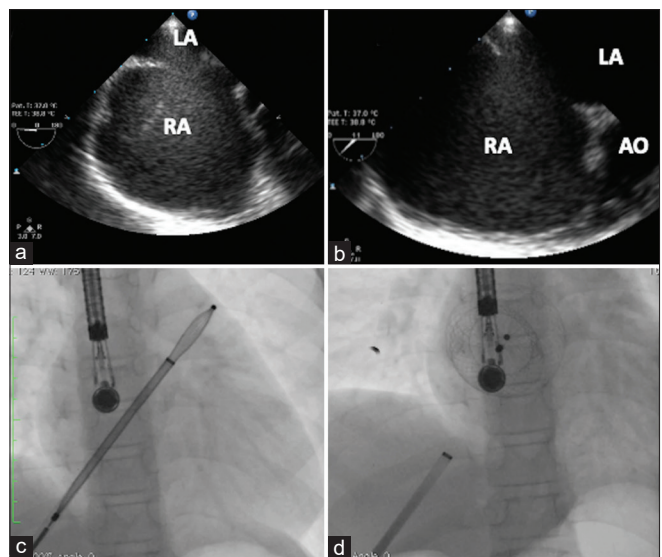


Figure 2: (a and b) TEE images of case 1 at 0° and 45° showing large FOASD with deficient retroaortic rim. (c) Plain fluoroscopy image in AP view showing 10-Fr flexor sheath (Cook Medical, Bloomington, MN, US) positioned in the LUPV with 20-mm Amplatzer septal occluder (Abbott medical MN, US). (d) Fluoroscopy image in anteroposterior view showing deployed Amplatzer septal occluder. TEE - Transesophageal echocardiography, FOASD - Fossa ovalis atrial septal defect, LUPV - Left upper pulmonary vein, RA - Right atrium, LA - Left atrium, AP - Anterio-posterior view

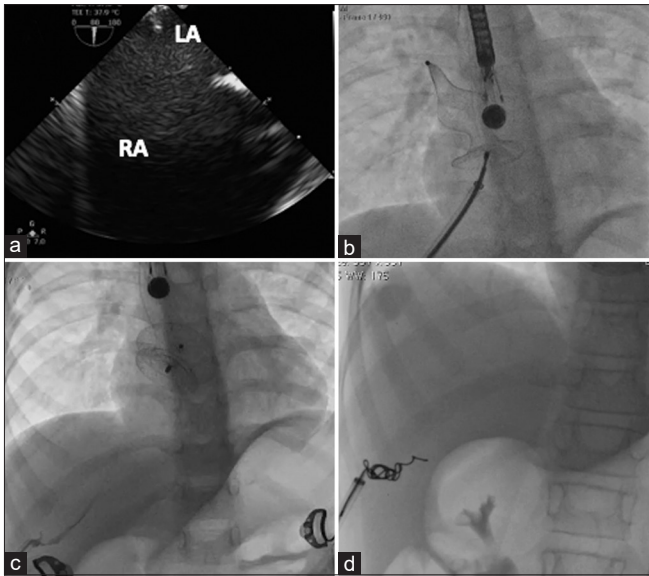


Figure 3: (a) TEE image of case 2 in 90° showing large FOASD with deficient inferior-posterior rim. (b) Plain fluoroscopy image in AP view showing deployment of ASO from RUPV. (c) Fluoroscopy image after deployment showing well-positioned device. (d) Closure of hepatic track using 0.035 stainless steel coils (Cook Medical, Bloomington, US). FOASD - Fossa ovalis atrial septal defect, ASO - Amplatzer septal occluder, RUPV - Right upper pulmonary vein, RA - Right atrium, LA - Left atrium, AP - Antero-posterior view

delivery sheath was withdrawn gradually till the distal hepatic vein and then to the outer quadrant of the liver parenchyma. The track in the hepatic parenchyma to subcutaneous tissue was closed using 0.035 inch stainless steel coils (Cook Medical, Bloomington, US) [Figure 3d]. Hemostasis was achieved and the patient was extubated. Ultrasound assessment was routinely done immediately and 4 h after the procedure to see any peritoneal collection in Morison’s pouch and pelvis. The assessment of liver function tests was done before discharge. Both the patients were discharged after 48 h and advised aspirin 3 mg/kg for 6 months.

DISCUSSION

Percutaneous transhepatic approach was initially done to perform portal vein interventions and to keep long-standing indwelling catheters.^[5] Later, Johnson *et al.* in 1995 performed cardiac catheterization and interventions through the transhepatic route.^[2] Shim *et al.* reported 30 transhepatic interventions in the pediatric population.^[3] Initially, procedures were performed under fluoroscopic guidance, but subsequently, ultrasound-guided approach became popular due to better identification of appropriate hepatic veins. This procedure can be done either on conscious patients under local anesthesia or intubated patients. Breath holding during the access time may avoid injury to the liver and access can be done predictably.

After an ultrasound assessment using a sector probe (to have more depth), a straight course hepatic vein should be identified for the access. A 20-G needle is used for the puncture to access the selected hepatic vein. Serial dilations of the hepatic tract should be done to prevent injury to the liver. This approach reduces complications such as intraperitoneal bleed, hepatic injury, and bile duct injury.^[3,6]

IVC interruption is one of the associations seen in several congenital heart defects. Preprocedural evaluation of this entity is important for the planning of the cardiac catheterization procedure. Alternative methods including transjugular or transhepatic approach may be useful in the presence of IVC interruption. Adwani *et al.* reported the ability of radiofrequency ablation through transhepatic approach in post-Fontan children.^[7] Later, several others published their data on transhepatic branch pulmonary artery interventions in postoperative cases of congenital heart diseases.^[8,9]

Emmel *et al.* performed FOASD device closure in a 3-year-old child with interrupted IVC.^[4] The transhepatic approach facilitates direct access to the atrial septum and left atrium. Larger sheaths can be used even in smaller children safely like in our second case. In the transhepatic approach, delivery sheath direction is perpendicular to the interatrial septum, facilitating alignment of the device.^[10] Our second case had small inferior-posterior rim and found difficulty in positioning the device by standard method, but we overcame this problem after deployment from RUPV. In the transjugular approach, either modification of sheath or the placement of guidewire in the left lower pulmonary vein or left ventricle is needed. Therefore, the transhepatic approach appears superior for closing large FOASD in case of IVC interruption.^[4] We successfully performed transcatheter device closure on two patients with large FOASD and deficient rims through percutaneous transhepatic approach. We chose flexor sheaths for the delivery of occluder as the alignment from the hepatic vein to the left atrium was straight.

CONCLUSIONS

Transhepatic approach is a promising alternative method for device closure of FOASD whenever femoral access is not possible. This method appears reasonably safe and allows larger sheaths even in smaller children.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patients have given their consent for images and other clinical information to be reported in the journal. The patients understand that their names and initials will

not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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