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## Case Report

## Percutaneous atrial septal defect closure through femoral and transjugular approaches in patients with interrupted inferior vena cava



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## ABSTRACT

Femoral venous approach is the classic route of percutaneous atrial septal defect (ASD) closure. But in patients with interrupted inferior vena cava (IVC) with azygos continuation the normal connection is lost rendering transcatheter intervention more complicated and innovative approaches should be sought. Transhepatic approach has been used with success but this approach is technically demanding and could lead to major complications. We report successful percutaneous secundum ASD closure in 2 patients with interrupted IVC. Transjugular route was used to close the secundum ASD in a 15-month-old girl. Percutaneous ASD closure through femoral venous route was performed in a 49-year-old woman. We found that both these approaches of percutaneous ASD device occlusion can be performed with success and safety.

**<Learning objective:** Interrupted inferior vena cava with azygos or hemiazygos continuation is a rare congenital anomaly but potentially complicates the usual percutaneous atrial septal defect (ASD) closure and therefore merits an accurate diagnosis before the procedure. Percutaneous ASD device closure can be performed with success and safety through femoral and transjugular approaches in patients with ostium secundum ASD who present with interrupted inferior vena cava.>

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## Introduction

Atrial septal defects (ASDs) account for approximately 6%–10% of congenital heart defects, with an incidence of 1 in 1500 live births [1] and a 97% probability of survival into adulthood [2]. Ostium secundum ASD accounts for approximately 75% of these defects with a female predominance of approximately 2:1. Patients with ostium secundum ASD usually remain asymptomatic during childhood and adolescence but may become symptomatic in infancy in case of large ASD and most commonly from the third to the fourth decade of life. Life expectancy and quality are reduced due to pulmonary circulatory overload, right heart failure, and pulmonary arterial hypertension, even Eisenmenger syndrome [3]. Percutaneous device closure is currently the treatment of choice. It provides similar efficacy and hemodynamic benefits with reduced complication rates and duration of hospital stay compared

to surgical closure [4]. The standardized percutaneous ASD closure procedure is performed through inferior vena cava (IVC) with femoral approach. However, in patients with interrupted IVC and azygos/hemiazygos continuation, this usual navigation is lost rendering the procedure difficult or impossible. Transhepatic approach for ASD device closure has been introduced with success but this route is technically challenging and could lead to major complications [5,6]. There have been some case reports of successful ASD device closure via femoral and transjugular approaches in this group of patients [7–9]. In this paper, we report successful percutaneous secundum ASD closure in two patients with interrupted IVC via femoral and transjugular routes in an adult and in a child.

## Case report

## Case 1

A 49-year-old female patient was referred to University Medical Center at Ho Chi Minh City for ASD closure. The presence of an ASD had been known for decades but was left untreated until recent

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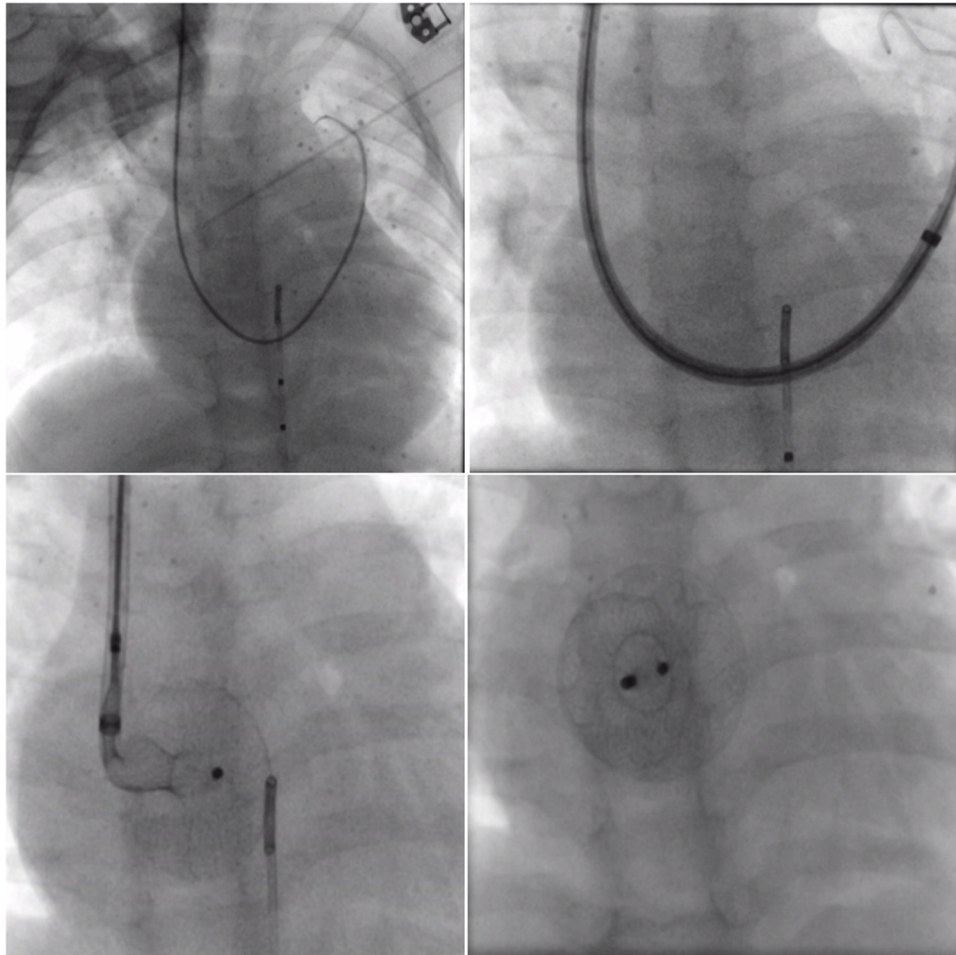


**Fig. 1.** Percutaneous secundum atrial septal defect closure through femoral approach in a 49-year-old female patient.

increasing exertional dyspnea. On clinical examination a soft systolic murmur was noted along the left upper sternal border. On transthoracic echocardiographic (TTE) examination a secundum ASD with diameter measured of 18 mm and sufficient rims was detected and these findings were confirmed by transesophageal echocardiography. In addition, a normal atrial situs, interrupted IVC with azygos continuation were also noted. On venography, a large azygos vein draining to the right-sided superior vena cava was seen. A catheter was first inserted to right femoral vein as usual and was advanced through the azygos vein up to the superior vena cava then curved to the right atrium. From the right atrium a hydrophilic wire and 5F Judkins right (JR) catheter were advanced to the left atrium and to left upper pulmonary vein (LUPV) as routine. Due to the sharp curve at the junction of azygos vein and the superior vena cava, we failed many times to exchange an Amplatzer guidewire through 5F JR. We decided to replace the 5F JR with a 6F JR. With better backup of 6F JR we could finally advance Amplatzer guidewire to LUPV. A 24-mm sizing balloon (NuMED, Hopkinton, NY, USA) was then inserted and inflated and the balloon-sized diameter of ASD was of 24 mm. In view of the unusual route with a sharp loop, we decided to oversize the device. An 11F delivery system was advanced to LUPV as usual (Fig. 1; top left), then a 26 mm AMPLATZER™ atrial septal occluder (St. Jude Medical, Inc., St Paul, MN, USA) was deployed from LUPV under fluoroscopic guidance (Fig. 1; top right). The position of the device was checked by TTE. The device was then successfully released as routinely performed (Fig. 1; bottom left and right).

#### Case 2

Next, we report a successful secundum ASD device closure through transjugular approach in a 9.5 kg, 15-month-old girl with interrupted IVC by using a “modified” delivery system namely PDA 7F (St. Jude Medical, Inc.) instead of ASD 7F. The child was referred to us for ASD closure. Clinical examination showed good physical development but she presented with exertional dyspnea. TTE showed a normal atrial situs, interrupted IVC, and azygos continuation with a secundum ASD of 11 mm of diameter with sufficient rims. Due to the interrupted IVC, we decided to puncture the right internal jugular vein. The right heart catheterization was performed with pulmonary pressures in normal range. A catheter 5F multipurpose (MP) catheter over a 0.035” hydrophilic guidewire was advanced until the LUPV was reached (Fig. 2; top left). The hydrophilic guidewire was exchanged with a stiff, 0.035” Amplatzer guidewire. An 18-mm sizing balloon (NuMED) was advanced over the stiff guidewire to ASD and inflated. The balloon-sized ASD diameter was of 13 mm. Given the fact that we were dealing with an unusual route, we decided to oversize the device. For a suitable curvature we used a 7F PDA delivery system instead of ASD delivery system (Fig. 2; top right). A 15 mm Amplatzer® septal occluder was positioned under fluoroscopic guidance (Fig. 2; bottom left) and was checked by TTE. The device was finally released successfully. The procedure was accomplished without any major complication (Fig. 2; bottom right).



**Fig. 2.** Percutaneous secundum atrial septal defect closure through transjugular approach in a 15-month-old girl.

## Discussion

Percutaneous transcatheter closure of a secundum ASD that was first reported by Mills and King in 1976 is now considered to be the treatment of choice for secundum ASDs [9]. The standardized percutaneous ASD closure procedure is performed through IVC with femoral approach. In heterotaxia, especially left isomerism, the IVC is absent and blood from the lower part of the body is drained through an azygos or hemiazygos vein. These veins are not connected directly to the right atrium but join the superior vena cava creating an acute angle before returning to the right atrium. Due to a longer and more tortuous trajectory of azygos vein, navigation to LUPV in the presence of a secundum ASD is more difficult than in patients with normal connection from IVC to right atrium. Due to this issue, the percutaneous device closure of an ASD in patients with interrupted IVC has been considered more difficult and challenging.

Transhepatic approach for ASD device closure has been introduced as an innovative technique [5,6]. However, this approach is technically demanding and not immune to complications. Major complications of the transhepatic approach include retro- or intraperitoneal bleeding, hemobilia, perforation of the gall bladder, portal vein thrombosis, pneumothorax, pleural effusions, and liver abscess or peritonitis and liver injury [9]. The femoral approach [7,8] and transjugular venous approach [9] have been used to occlude secundum ASD. The number of cases are, however, still limited. In this communication, we report two

successful percutaneous ASD closures in 2 patients with interrupted IVC using transjugular and femoral approach.

For the first case where femoral approach was performed, we did encounter difficulty in advancing the stiff guidewire, an essential step for safely introducing the delivery system, when using the 5F JR catheter. Replacing the 5F catheter with a 6F catheter over 260 cm exchange guidewire might render adequate support required for stiff Amplatzer guidewire to reach LUPV. This is a practical point that may help the interventionist in coping with the same difficulty as we did.

Our second case may be one of the smallest patients who underwent transjugular ASD device closure. In this case, for better conforming to a sharp angle, we decided to use C-shape delivery system instead of obtuse end catheter so that its distal end can be advanced into left atrium and then to LUPV much easier.

Both procedures were achieved in a good duration and without complication. In the first case, where femoral route was used, the patient preparation, site of puncture, and the running of procedure were not different from standardized usual procedure and the patient was fully awake. However, the navigation through sharp curve at the junction of azygos vein and the superior vena cava was sophisticated and required an experienced interventionist. In the second case, where the transjugular approach was used, the preparation was at first a bit different but the navigation to right atrium was much easier. With very limited experience from these two cases, we would like to recommend the transjugular approach for ASD device closure in patients with interrupted IVC.

**In conclusion**, percutaneous ASD closure in patients with interrupted IVC and azygos continuation through femoral and transjugular approaches are feasible and safe in both adult and pediatric population. Accurate preprocedural diagnosis, detailed planning, and innovative techniques are paramount for safe and successful intervention.

### Conflict of interest

The authors declare that there is no conflict of interest.

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