THE ANATOLIAN JOURNAL OF CARDIOLOGY

Official journal of the TURKISH SOCIETY OF CARDIOLOGY

Successful Transjugular Percutaneous Atrial Septal Defect Closure in a Dextrocardia Patient with Kartagener's Syndrome, Situs Inversus, Interrupted Inferior Vena Cava, and Bilateral Superior Vena Cava

CASE REPORT

INTRODUCTION

Transcatheter device closure of a secundum atrial septal defect (ASD) is now a well-accepted treatment alternative to surgical intervention, and it is usually carried out via the inferior vena cava (IVC) using the femoral venous (FV) approach.[1] In patients who have an interrupted IVC, the procedure can be accomplished by a femoral, transhepatic, or transjugular route.[2,3] However, in patients with interrupted IVC, which drains into the coronary sinus (CS), it is impossible to close the defect percutaneously via the femoral approach. We report a challenging case with Kartagener's syndrome, dextrocardia with situs inversus, interrupted IVC with azygos continuation, which drains into the CS, and bilateral superior vena cava (SVC). A successful transjugular percutaneous secundum ASD closure was accomplished in this case.

CASE REPORT

A 10-year-old male patient was referred to our department for percutaneous closure of a secundum-type ASD. Transesophageal echocardiography (TEE) indicated a 14.6 mm interatrial septal defect obtained from the maximal color-flow jet width (Figure 1A and B, Supplementary Video 1). Under TEE guidance, contrast injection revealed that the IVC was interrupted with communication through the right SVC by the azygos vein, which subsequently continued with the CS (Figure 2A, Supplementary Video 2). The procedure was performed through the left internal jugular vein (IJV). We did not convert the scopy from the left to the right, but the procedure was performed on the left side of the patient. A 6-French JR4 catheter was advanced via the left IJV and positioned in the left ventricle (LV) through the right atrium (RA) and the left atrium using a 0.035" hydrophilic guidewire. The hydrophilic wire was replaced with an extra stiff 0.035" wire, and the wire tip was embedded in the LV. With TEE guidance, it was decided that a 16 mm Amplatzer™ septal occluder (Abbott, Abbott Park, Ill, USA) was the appropriate device to perform the closure procedure. The 7-French long sheath was advanced over the extra stiff guidewire and placed into the LV to keep the guidewire and long sheath in greater support and to facilitate the intervention (Figure 2B). The long sheath was then carefully pulled into the LA while advancing the device. The LA disk of the device was deployed in the LA; then, the system was progressively pulled back while holding the long sheath fixed. Finally, the RA disk was deployed. Subsequent TEE imaging revealed an ideal location for the device, and the device was completely released (Figure 1C and 2C, Supplementary Video 3). The hemostasis was achieved by manual compression and the procedure was finished uneventfully. The patient was discharged with acetylsalicylic acid therapy.

DISCUSSION

In patients with secundum-type ASD and interrupted IVC, a careful assessment of alternate accessible sites is needed and the transcatheter ASD closure can be performed by the FV.[4] However, in our case, the procedure via either the FV or the

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Cite this article as: Yücel İK, Aslan Çınar A, Sürücü M, Çelebi A. Successful transjugular percutaneous atrial septal defect closure in a dextrocardia patient with kartagener's syndrome, situs inversus, interrupted inferior vena cava, and bilateral superior vena cava. *Anatol J Cardiol.* 2023;27(1):47-49.

DOI:10.14744/AnatolJCardiol.2022.2065

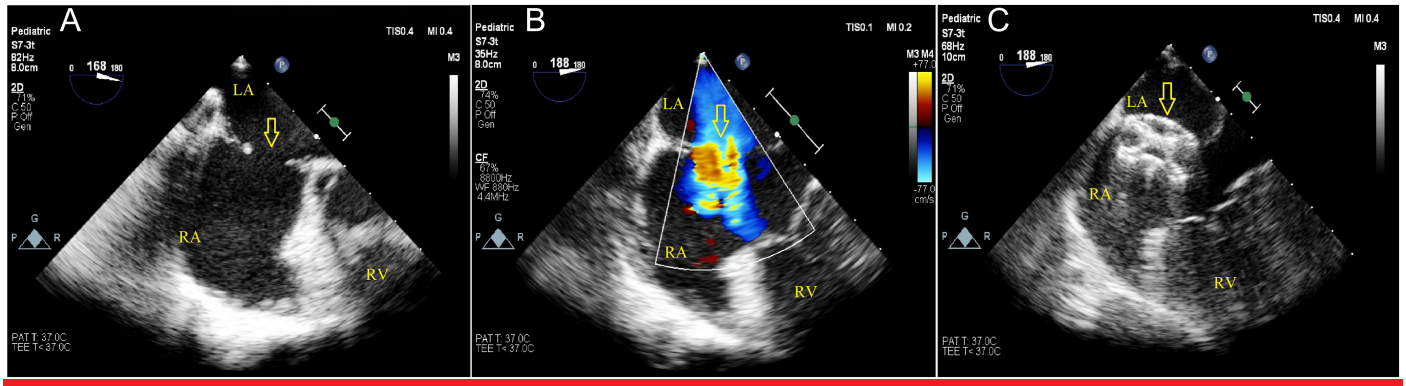

Figure 1. (A) Transesophageal echocardiography 2-dimensional image showing an interatrial septal defect obtained. (B) Transesophageal echocardiography image showing an interatrial septal defect obtained from the maximal color-flow jet width. (C) Transesophageal echocardiography image showing successful deployment of the Amplatzer septal occluder.

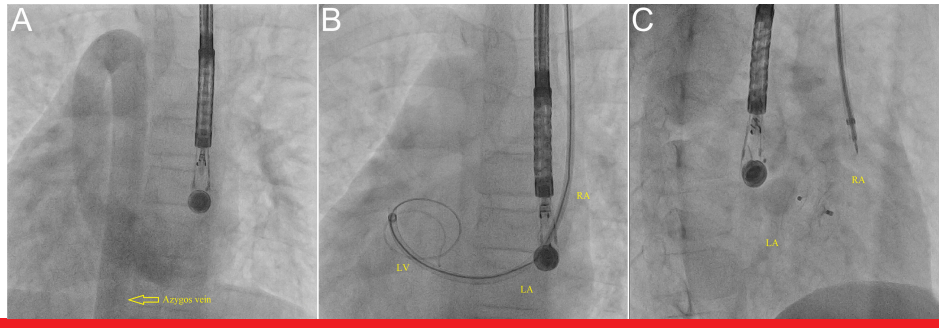

Figure 2. (A) Fluoroscopy image showing a communication through the right superior vena cava by the azygos vein, which subsequently continued with the coronary sinus. (B) Fluoroscopy image showing the placement of a 7-French long sheath into the left ventricle to keep it in greater support. C) Fluoroscopy image showing successful deployment of the Amplatzer septal occluder.

right IJV was not possible because the azygos vein drained into the CS. Furthermore, due to its ability to accommodate significantly larger sheaths and its optimal alignment to the atrial septum, left IJV access was considered the most suitable approach.

Until now, secundum-type ASDs have been occluded with different routes in a few patients with interrupted IVC and different types of implanted devices.[5,6] Table 1 presents a short description of similar cases in the literature with all the necessary procedure details.

The most difficult parts of ASD closure via the transjugular approach are crossing the septal defect with an extra stiff guidewire and inserting an extra stiff guidewire into the pulmonary vein to carry the delivery system. Another challenge of the transjugular venous approach is ensuring that the device is correctly oriented and aligned with the atrial septum to avoid protrusion in the LA. Furthermore, this technical challenge is even more complicated in situations of dextrocardia with situs inversus and an interrupted IVC, making the procedure considerably more difficult.[6,7] As demonstrated in our case, keeping the long sheath in the LV offered greater support and facilitated the intervention. However, in this method, the long sheath should be pulled into the LA just before deploying the LA disk in order not to damage the mitral valve and chordae. We also saw several benefits of the new Abbott delivery cable in this procedure. The delivery system's highly flexible wire tip made it easier

Table 1. A Brief Outline of All Interrupted IVC and Secundum ASD Cases with All the Relevant Procedure Information

Age	Gender	Access Side	Device Diameter	Device Type
8 years old	Male	Right IJV	10 mm	Amplatzer septal occluder
29 years old	Male	Transhepatic vein	10 mm	Amplatzer septal occluder
49 years old	Female	Femoral vein	26 mm	Amplatzer septal occluder
15 months	Female	Right IJV	15 mm	Amplatzer septal occluder
3 years old	Male	Left IJV	19 mm	Amplatzer septal occluder
12 years old	Female	IJV	24 mm	Ceraflex septal occluder

ASD, atrial septal defect; IJV, internal jugular vein; IVC, inferior vena cava.

for the distal end of the cable to bend more than 90°, which facilitated the device's alignment with the atrial septum during implantation.

CONCLUSION

As shown in our case, we believe that the inability to use standard IVC access should not dismiss percutaneous ASD closure as an optimal treatment choice in patients with secundum ASD and interrupted IVC.

Informed Consent: Informed consent was obtained from the patient's legal guardians.

Supplementary Video 1: Video showing an interatrial septal defect obtained from the maximal color-flow jet width.

Supplementary Video 2: Video showing a communication through the right superior vena cava by the azygos vein, which subsequently continued with the coronary sinus.

Supplementary Video 3: Video image showing successful deployment of the Amplatzer septal occluder.

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