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Does Transcatheter Therapy Replace Surgery? Transcatheter Closure of Sinus Venosus Atrial Septal Defect and Partial Pulmonary Venous Return

# INTRODUCTION

Sinus venosus atrial septal defect (SVASD) occurs due to the lack of development of the common wall between the superior vena cava (SVC) and the right upper pulmonary vein (RUPV). So the RUPV drains into the SVC.<sup>1</sup> In recent years, transcatheter closure has been an alternative to surgery in selected cases.<sup>2</sup> Herein, we shared our experience of treating a case with a step-by-step approach.

# **CASE REPORT**

A 49-year-old female patient had occasional palpitations. An adult cardiologist diagnosed SVASD by echocardiography and referred the patient to a cardiovascular surgery clinic. In our echocardiographic examination, the patient had additional partial anomalous pulmonary venous return (PAPVR) in addition to large SVASD. Cardiac computed tomography (CT) angiography was performed to better elucidate the cardiac anatomy and evaluate the suitability for transcatheter closure. Computed tomography angiography revealed the following: right lung upper lobe and middle lobe lateral segment pulmonary veins drained to the right SVC,  $13 \times 20$  mm SVASD, enlargement of the right heart chambers, and persistent left SVC without intercaval connection (Video 1). The patient seemed suitable for transcatheter SVASD closure.

Under general anesthesia, 10F sheaths were introduced into the right femoral vein (RFV) and 6F sheath into the RFV and right internal jugular vein (RIJV), and then 5000 IU intravenous heparin was given. The defect and the pulmonary veins were evaluated with transesophageal echocardiography (TEE) (Video 2).

- A venovenous (VV) loop was established between RFV and RIJV with a 0.035" stiff wire.
- A TEE-guided transseptal puncture was performed, and then a 5F pigtail catheter was placed into the RUPV.
- Contrast injections into the pulmonary vein and right SVC revealed that RUPV was draining into the right SVC, and the distal diameter of the SVC was 18 mm. Since there was no intercaval connection in the patient, left SVC evaluation was not required, but the drainage of the azygos system to the right SVC was evaluated.
- A 18 × 50 mm VACS-2 balloon (Osypka Medical Inc., San Diego, CA, USA) was advanced over the VV loop covering SVASD and PAPVR for test occlusion. The balloon was inflated, and the following evaluations were performed.
- Contrast injection to the right SVC revealed that the azygos vein was non-occluded.
- Transesophageal echocardiography showed following;
- Two-Dimensional echo revealed complete closure of the atrial septal defect.
- Color Doppler revealed no stenosis of RUPV.
- Continuous wave Doppler revealed no gradient between RUPV and the left atrium (LA).



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## **CASE REPORT**





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- There was no increase in RUPV pressure, and there was no pressure gradient between RUPV and LA.
- Contrast injection into the RUPV revealed normal drainage into the left atrium without stenosis.
- The same evaluations were then repeated with a 22-mm non-compliant balloon, and the same results were found.

For the appropriate stent selection, a marker pigtail catheter was advanced over the VV loop and the stent length was estimated with contrast injections. Since it is difficult to find 8-10-cm-long covered stents in our country, it was decided to close the defect with the stent-in-stent method.

Initially, a 48 mm bare Optimus XL stent (AndraTec GmbH) was crimped on the  $22 \times 50$  mm balloon in balloon catheter (BIB) (Numed, Hopkinton, NY, USA) and implanted in the SVC side of the defect. This stent would both make the landing zone for the second stent and ensure that the flow of the azygos system would not be occluded. Then, a 57 mm covered Optimus XL stent crimped on a  $22 \times 50$  mm BIB catheter was advanced into the bare stent and implanted in the distal part of the defect. Here, attention was paid to ensure that the covered stent was 2 cm caudal to the first stent, covering the defect, and protruded 2 cm into the right atrium (Video 3).

There was no residual shunt in SVASD in control injections or TEE controls. RUPV was completely draining into the left atrium, and there was no stenosis here. In the echocardiographic controls performed the day after the procedure, there was no stenosis in the pulmonary vein and no residual shunt through the ASD. In addition, no fracture was detected in the stent in the telecardiography. The electrocardiogram was in sinus rhythm. The patient was discharged for outpatient follow-up.

### DISCUSSION

In the treatment of SVASD, it was previously thought that the only way to repair this defect was surgery. In 2013, Abdullah et al<sup>3</sup> described transcatheter SVASD closure. Subsequently, several case series have been published, and the technique has improved over time. The basic idea is to close the defect by advancing a covered stent with a diameter that will not prevent pulmonary vein flow.<sup>4</sup> Transesophageal echocardiography provides information about the diameter of the defect, its caudal extension to the fossa ovalis, and pulmonary vein openings and flows. More precise data on the anatomy of the pulmonary veins and their relationship to the defect can be obtained with cardiac CT and/or magnetic resonance imaging. Preparation for the procedure can be done by simulating cardiac anatomy with a 3-dimensional printer.<sup>5</sup> In cases where standard stents are not appropriate, custommade stents can be produced in light of all these data.<sup>6</sup> In countries where access to custom-made stents is difficult, the procedure can be performed with 2 or more stents placed inside each other.7 We also used 2 stents, 1 bare and 1 covered, in our patient. Sivakumar et al<sup>8</sup> showed that appropriate patient selection can be made by TEE and angiographic balloon tests without the need for advanced imaging techniques. While it was previously thought that this procedure could only be applied to adult patients, in recent years, the procedure has also been successfully applied to pediatric patients that have a SVC diameter over 18 mm.<sup>8</sup>

#### CONCLUSION

Transcatheter SVASD closure is an alternative procedure to surgery for selected patients. Short-term follow-ups regarding this still-new technique are promising. The technical details of the process are under development.

**Informed Consent:** Written informed consent was obtained from the patient for the publication of this case report and accompanying videos.

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**Video 1:** 3D constructed CT images: The anatomy of the sinus venosus defect and its relationship with the anomalous draining pulmonary veins.

Video 2: Consecutive TEE images taken during the procedure.

Video 3: Consecutive angiographic images taken during the procedure.

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