

Transcatheter Closure of Ruptured Sinus of Valsalva Aneurysm

with Double-Disc Perimembranous VSD Occluder in
Man with Mechanical Aortic Valve

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Sinus of Valsalva aneurysm, a rare cardiac anomaly, can be life-threatening if it ruptures. Transcatheter closure has emerged as an effective alternative to surgical management; however, it has rarely been reported in patients with previous mechanical aortic valve replacements. We describe the case of a 45-year-old man who presented with a ruptured aneurysm of the noncoronary sinus of Valsalva 14 years after the implantation of a mechanical aortic valve. The ruptured aneurysm was closed by transcatheter means with use of a double-disc perimembranous ventricular septal defect occluder. The patient remained asymptomatic one year after the procedure. Our case suggests that transcatheter closure with use of this type of occluder is a viable method for successfully repairing ruptured sinus of Valsalva aneurysms in patients who have mechanical aortic valves. (Tex Heart Inst J 2019;46(3):211-4)

Key words: Aneurysm, ruptured/pathology/therapy; balloon occlusion/instrumentation; cardiac surgical procedures/methods; heart aneurysm/therapy; heart valve prosthesis implantation/methods; septal occluder device; sinus of Valsalva/pathology; treatment outcome

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Ruptured sinus of Valsalva aneurysm (SVA) is a rare cardiac condition, with a higher incidence in Asian populations than in Western populations.¹ Although SVA can develop consequent to endocarditis, trauma, or degenerative processes, it is typically a congenital condition caused by a weakness of the wall of the sinus of Valsalva, which leads to aneurysmal formation.²⁻⁴ Rupture of the aneurysm can present with a variety of symptoms; presentations range from asymptomatic to cardiogenic shock.³⁻⁶ Surgical repair has been the main treatment for ruptured SVA. Transcatheter closure (TCC) has recently emerged as an alternative, less invasive treatment for this patient population.⁵⁻⁸ However, TCC of ruptured SVA may be technically challenging in patients who have undergone mechanical aortic valve replacement (AVR). We describe the case of a 45-year-old man who presented with symptomatic heart failure secondary to a ruptured aneurysm of the noncoronary sinus of Valsalva (NCS) 14 years after mechanical AVR. We performed TCC of the ruptured aneurysm with use of a Chinese double-disc perimembranous A4B2 ventricular septal defect (VSD) occluder (Lifetech Scientific [Shenzhen] Co., Ltd.), which is similar to the AMPLATZER™ VSD occluder (St. Jude Medical, part of Abbott). To our knowledge, this is the first report of TCC of a ruptured SVA with use of this A4B2 occluder in a patient with a mechanical aortic valve. We discuss how this procedure enables treatment for carefully selected patients who present with ruptured SVA after AVR.

Case Report

In December 2016, a 45-year-old man was admitted to our hospital with a 3-month history of progressive exertional dyspnea. Fourteen years earlier, he had undergone AVR with implantation of a mechanical prosthetic valve to treat his severe aortic stenosis. Cardiac examination revealed a continuous murmur audible at the left sternal border. Transthoracic echocardiograms (TTE) showed an aneurysm of the NCS protruding into the right ventricular outflow tract (RVOT), with the characteristic windsock appearance. Color-flow images showed turbulence across the ruptured aneurysm (Fig. 1). The prosthetic valve was in good condition, and there was no evidence of paravalvular leak. Results of cardiac catheterization included a pulmonary artery pressure of 51/25 mmHg (mean, 33 mmHg) and an aortic pressure of 130/64 mmHg (mean, 86 mmHg). Oximetry studies detected left-to-right shunting

at the ventricular level (calculated Qp/Qs ratio, 1.6:1). Ascending aortograms confirmed a rupture of the NCS aneurysm into the RVOT (Fig. 2A). The morphologic features were like those of an aneurysm-shaped VSD; multiple outlets (holes) were present in the right side of

the ruptured SVA, the narrowest diameter of which was approximately 6.5 mm.

We decided to use a TCC approach to repair the aneurysm. The defect was crossed from the aortic side with use of a 6F multipurpose catheter and a 0.035-in, 260-cm hydrophilic exchange guidewire (Terumo Medical), which was then snared with an Amplatz gooseneck snare (Microvena Corp.) from the pulmonary artery and exteriorized from the femoral vein. A stable arteriovenous wire loop was thus established through the ruptured SVA, over which an 8F delivery sheath was advanced from the femoral vein and placed into the ascending aorta across the defect. A 12-mm muscular VSD occluder (Lifetech Scientific) was deployed through the long sheath under fluoroscopic guidance. However, aortograms showed that the prosthetic valve prevented the occluder from maintaining its optimal configuration and led to substantial residual shunting. The device was retrieved, and we placed a 10–12-mm duct occluder (Lifetech Scientific). However, residual shunting was still substantial. Subsequently, we deployed a 10-mm A4B2 double-disc perimembranous VSD occluder (Lifetech Scientific). This device has a symmetric left disc (diameter, 8 mm larger than the waist cylinder) and a right disc 4 mm larger than the cylinder. The waist cylinder is 2.5 mm long.⁹ Aortograms confirmed that the device was in a good position, without substantial residual shunting or aortic valve regurgitation (Fig. 2B). The occluder was then completely released (Fig. 2C). The next day, TTE showed that the occluder remained in a stable position, with no RVOT obstruction (Fig. 3). The patient recovered uneventfully, was discharged from the hospital 3 days later, and was prescribed lifelong warfarin therapy. At his one-year follow-up examination, he was asymptomatic; a TTE and a cardiac computed tomographic angiogram (Fig. 4) showed a well-seated closure device without substantial residual shunting.

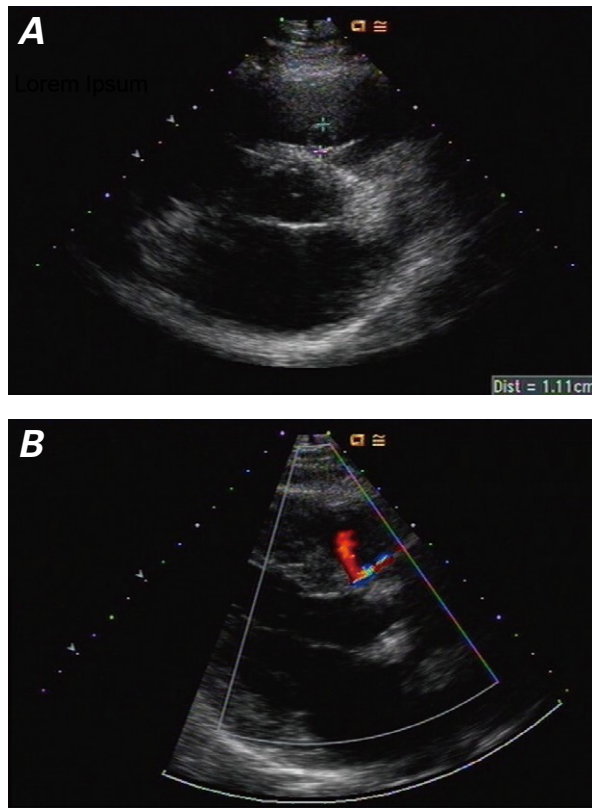


Fig. 1 Transthoracic echocardiograms show **A**) a ruptured aneurysm of the noncoronary sinus into the right ventricular outflow tract (parasternal short-axis view) with **B**) turbulent color flow across the ruptured aneurysm (parasternal long-axis view).

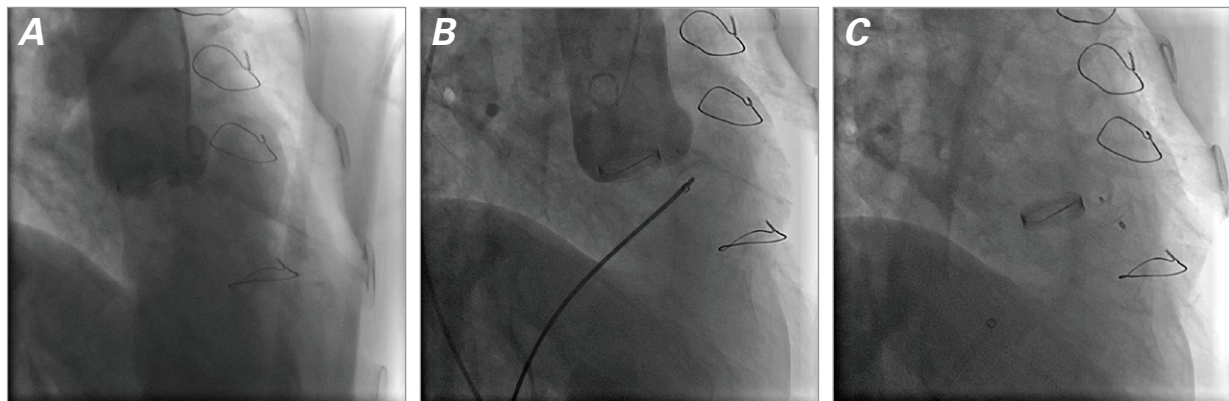


Fig. 2 **A**) Aortogram (caudal right anterior oblique view) shows the aneurysm of the noncoronary sinus ruptured into the right ventricular outflow tract. **B**) Repeat aortogram shows occlusion of the aneurysm with use of the A4B2 ventricular septal defect occluder. **C**) The device was released in good position.

Supplemental motion images are available for Figs. 2A and 2B.

Discussion

Ruptured SVA, which constitutes less than 1% of all congenital cardiac anomalies that necessitate surgical treatment,¹ can cause profound hemodynamic changes. This anomaly is typically caused by a congenital deficiency of elastic and muscular tissue in the aortic sinus, but it can also result from endocarditis, surgery, or trauma. Aneurysmal formation and progressive dilation (which in our patient took an uncommonly long 14 years) can culminate in aneurysm rupture into the cardiac chambers.^{2-4,6} In our patient's case, no ruptured SVA had been found before the AVR operation or during surgical exploration. Therefore, we postulated that his SVA was primarily caused by the absence of the elastic lamellae of the aortic media, which would weaken the aortic wall; his previous surgical AVR might have also contributed to aneurysm formation. The site of origin for SVAs is usually the right coronary sinus, followed by the NCS, with drainage typically into the right-sided heart chambers.^{1,6} Clinical presentation is determined chiefly by the magnitude of left-to-right shunting, the rapidity of aneurysm rupture, and the chamber into which the rupture drains. A large ruptured SVA with substantial left-to-right shunting can lead to acute congestive heart failure, cardiac tamponade, or sudden death.^{4,6}

Surgical repair has been the main treatment for ruptured SVAs; however, TCC has recently emerged as an effective alternative in carefully selected patients. Favorable immediate and long-term outcomes of TCC have increasingly been reported.^{5,8} Selecting a suitable occlusion device is a chief factor in determining the outcomes of TCC. Among several available devices, the AMPLATZER Duct Occluder is most frequently used, because it is suited to the morphologic characteristics of most ruptured SVAs, which typically present with a windsock-like structure with a broader aortic end.⁷ However, there is currently no standard method for selecting occluders. The individual anatomic characteristics of the ruptured SVA, including the size and location of the defect, the shape of the ruptured SVA, and the adjacent structures, guide the selection of the device.

Transcatheter closure of a ruptured SVA may be technically challenging in patients who have undergone AVR. Schaeffler and colleagues¹⁰ reported the case of a patient whose aneurysm of the right coronary sinus ruptured into the RVOT 4 years after AVR; the rupture was treated successfully by means of TCC with use of an AMPLATZER muscular VSD occluder. Zhong and co-authors⁷ described an aneurysm of the NCS that ruptured into the right ventricle in a 47-year-old woman who had undergone AVR 3 years earlier. She too underwent successful TCC of the ruptured aneurysm with use of an AMPLATZER muscular VSD occluder. To our knowledge, ours is the first report of TCC of a ruptured SVA with use of an A4B2 perimembranous

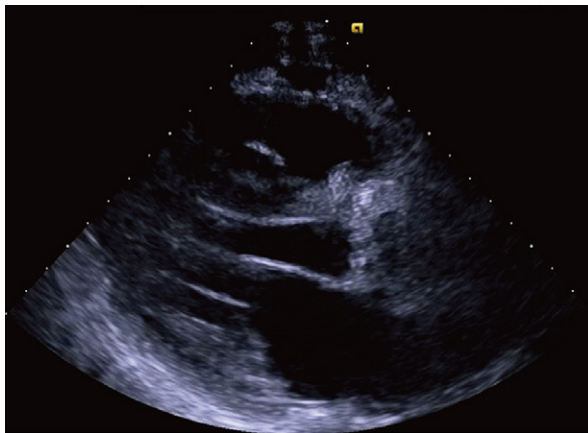


Fig. 3 Postprocedural transthoracic echocardiogram (parasternal long-axis view) shows stable occluder position and no right ventricular outflow tract obstruction.

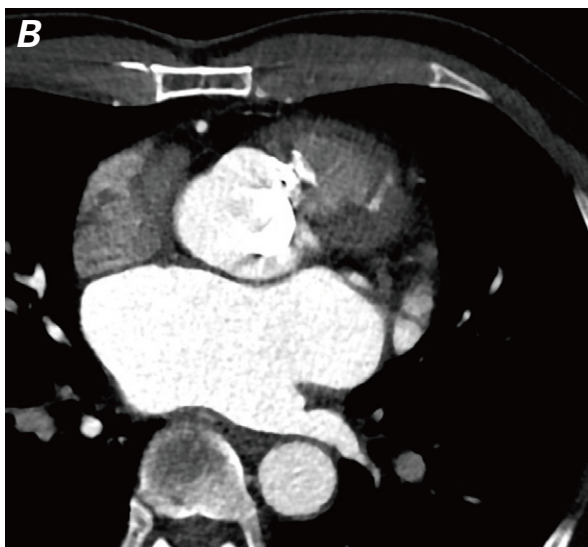
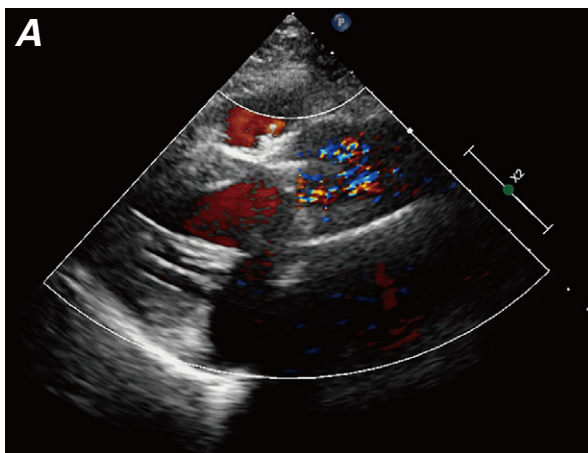


Fig. 4 At one year, **A**) a transthoracic echocardiogram (parasternal long-axis view) shows a well-seated closure device without substantial shunting, and **B**) a cardiac computed tomographic angiogram shows an intact occluder and resolution of the ruptured aneurysm.

VSD occluder in a patient who had a mechanical aortic valve. Several holes had formed over a large area in the right side of our patient's SVA. In view of these anatomic characteristics, the A4B2 VSD occluder was the most suitable device: its small waist matched the defect diameter so that the discs extended fully after occluder placement. The defect entrance and outlets were covered completely, verifying that one such occluder can close multiple holes simultaneously.¹¹ Moreover, when compared with the AMPLATZER Duct Occluder, the shorter waist length of the A4B2 VSD occluder prevented excessive protrusion of the device into the RVOT, thus avoiding iatrogenic RVOT obstruction.

Acknowledgment

We thank Sybille Redmond, Research Administrative Assistant, Minneapolis Heart Institute Foundation, Minneapolis, Minnesota, for her help in revising our manuscript.

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