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Successful Percutaneous Closure of an Aorto-right Ventricular Fistula After Sutureless Aortic Valve Replacement: A Case Report

Dikişsiz Aort Kapak Replasmanı Sonrasında Oluşan Aorta Sağ Ventrikül Fistülünün Perkütan Yolla Başarılı Bir Şekilde Kapatılması: Olgu Sunumu

ABSTRACT

Rupture of a sinus of valsalva aneurysm (SVA) and the development of an aorto-right ventricular fistula (ARVF) is a rare condition, associated with high morbidity and mortality rates if left untreated. Opening of the SVA rupture into the right heart chambers may result in various morbidities, such as pulmonary hypertension. We present a case of a patient who developed ARVF following sutureless aortic valve replacement, and was subsequently treated successfully via a percutaneous approach.

Keywords: Ruptured sinus of valsalva aneurysm, transcatheter closure, sutureless aortic valve replacement

ÖZET

Sinüs valsalva anevrizma (SVA) rüptürü ve sonrasında görülen aorta-sağ ventrikül fistülü (ASVF) nadir görülen bir durumdur ve tedavi edilmediği takdirde yüksek morbidite ve mortalite oranlarına sahiptir. SVA rüptürünün sağ kalp boşluklarına açılması pulmoner hipertansiyon ile sonuçlanabilir. Sutureless (dikişsiz) aort kapak replasmanı sonrasında ASVF gelişen ve perkütan yolla başarılı bir şekilde tedavi edilen bir olgu sunuyoruz.

Anahtar Kelimeler: Sinüs valsalva anevrizma rüptürü, trankatater kapatma, dikişssiz aort kapak replasmanı

CASE REPORT
OLGU SUNUMU

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The rupture of a sinus of Valsalva aneurysm (SVA) and the development of an aorto-right ventricular fistula (ARVF) constitute a very rare pathology, which can result in right ventricular overload and pulmonary hypertension if left untreated. A ruptured SVA has a poor prognosis, with high associated morbidity and mortality rates. Congenital SVA is often associated with Marfan syndrome or Ehlers-Danlos syndrome, while acquired aneurysms are linked to trauma, atherosclerosis, infective endocarditis, and iatrogenic injury during aortic valve replacement.[1] To date, several cases of SVA rupture after sutureless aortic valve replacement have been reported.[2,3] To the best of our knowledge, only one case of SVA rupture treated with percutaneous closure following sutureless aortic valve replacement has been documented.[2] Although the most commonly reported cases of ARVFs are secondary to the rupture of a congenital aneurysm of one of the sinuses of Valsalva, the development of an ARVF is an extremely rare complication following aortic valve replacement.[1-3] Herein, we report a case of successful percutaneous closure of an ARVF that developed subsequent to sutureless aortic valve replacement.

Case Report

A 72-year-old female patient, with a history of Perceval sutureless valve (LivaNova PLC, London, UK) replacement due to severe aortic stenosis, simultaneous right coronary artery bypass surgery, and a prior permanent DDDR pacemaker implantation for complete atrioventricular block, was admitted to our outpatient clinic presenting with dyspnea. Preoperatively, she exhibited no aneurysm of the aortic root or sinus

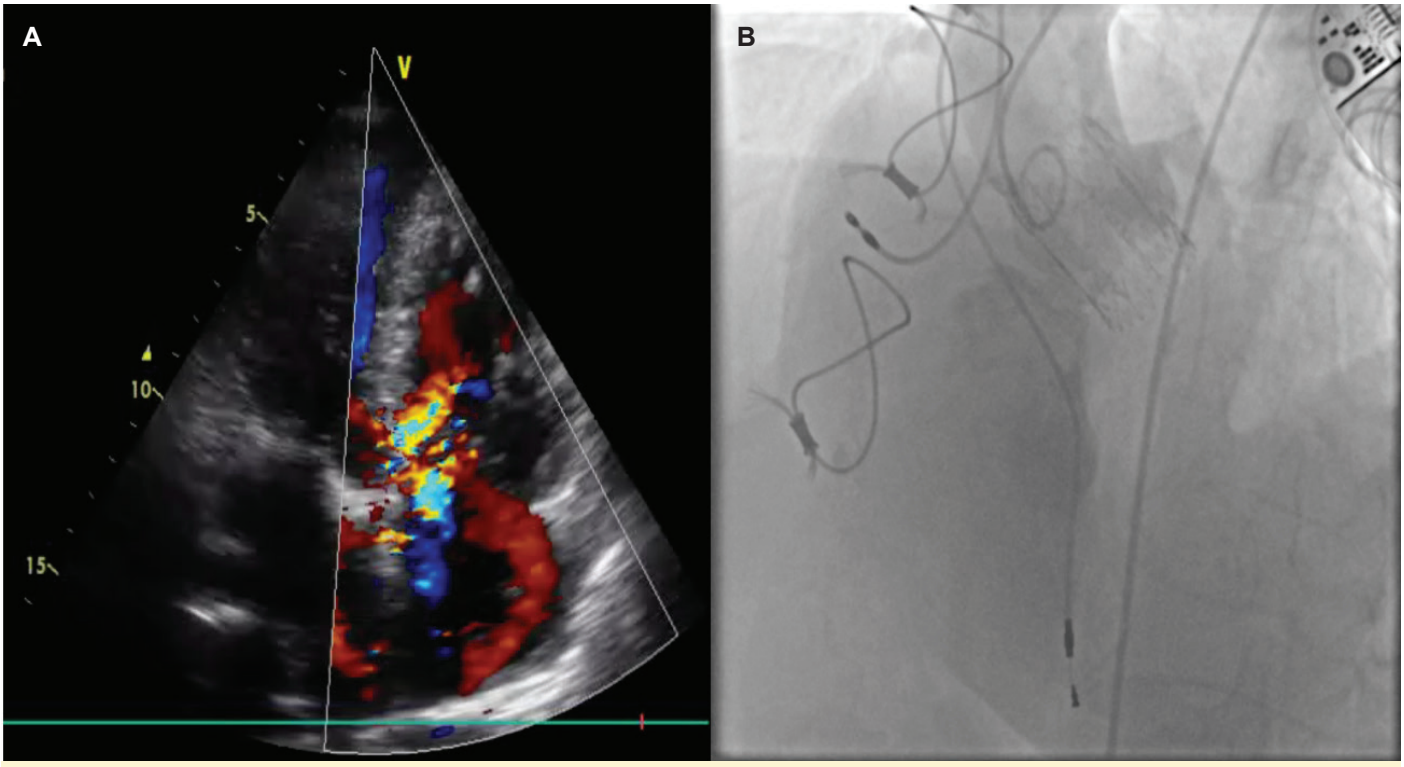

Figure 1. Color Doppler image depicting the aorto-right ventricular fistula (ARVF) extending from the aorta to the right ventricle (A), Aortic root angiogram illustrating the fistula's trajectory from the aorta to the right ventricle (B).

of Valsalva. Cardiac auscultation revealed a grade 4 pansystolic murmur over the apex. Transthoracic echocardiography (TTE) revealed a bioprosthetic aortic valve with a gradient of 32/19 mmHg, dilated right heart chambers, moderate tricuspid regurgitation, and an estimated systolic pulmonary artery pressure of 50 mmHg. Color Doppler imaging identified an ARVF extending from the aorta to the right ventricle (RV) (Figure 1A), with no evidence of infective endocarditis observed. Cardiac catheterization was performed, yielding a Qp/Qs ratio of 1.8, and ARVF was further confirmed through aortography imaging (Figure 1B). After evaluation by the heart team, the decision for surgical intervention was made. However, the patient opted for a percutaneous closure procedure instead. The femoral arterial and venous introducer sheaths were percutaneously inserted into the left femoral artery and the right femoral vein, respectively. Intravenous unfractionated heparin was administered prior to the procedure at a dose of 100 IU/kg. A 6F right guiding catheter (Judkins, Boston Scientific) was positioned within the Perceval valve, adjacent to the defect. Subsequently, a 0.035-inch hydrophilic wire (Roadrunner® PC Hydrophilic Wire Guide, Cook Medical) was threaded from the defect to the RV (Figure 2A). Following this, the guiding catheter, threaded over the hydrophilic wire, was advanced into the RV, and the hydrophilic wire was subsequently replaced with a superstiff wire (Amplatz Super Stiff™ Guidewire, Boston Scientific). The RV was accessed through the right femoral vein using a 6F right guiding catheter. The superstiff wire was captured with the help of a snare [Exeter Snare (macro), snare length: 125 cm, loop diameter: 20 mm, AndraTec Medical Devices], and an arterial-venous wire loop was established between the right femoral artery and the left femoral vein through the ruptured SVA (Figure 2B). A balloon-sizing procedure (Abbott Laboratories, Abbott Park, Illinois) was performed to determine the diameter of the defect, which was measured to be approximately 8 mm by aortography (Figure 2C). Under fluoroscopic guidance, the 10-8 mm AMPLATZER™ Duct Occluder device (Abbott Laboratories, Abbott Park, Illinois) was advanced through the venous route and inserted into the ruptured SVA, demonstrating no leakage into the RV (Figure 2D). Post-op TTE showed no change in aortic valve functions and no leakage from the aorta to the RV. A control aortography showed no signs of significant aortic regurgitation or compression of the coronary arteries. The patient was discharged two days after the procedure, having made a good recovery. Post-procedure, a 6-month regimen of acetylsalicylic acid (ASA) and clopidogrel was prescribed. At the 3-month follow-up, the patient was in good clinical condition, with no echocardiographic changes noted.

ABBREVIATIONS

ARVF	Aorto-right ventricular fistula
ASA	Acetylsalicylic acid
ICE	Intracardiac echocardiography
RV	Right ventricle
SVA	Sinus of Valsalva aneurysm
TTE	Transthoracic echocardiography

Discussion

SVA, a rare anomaly that can be either congenital or acquired, is more common in males with a ratio of 4:1.[4] SVA results from the weakness of the elastic lamina at the junction between

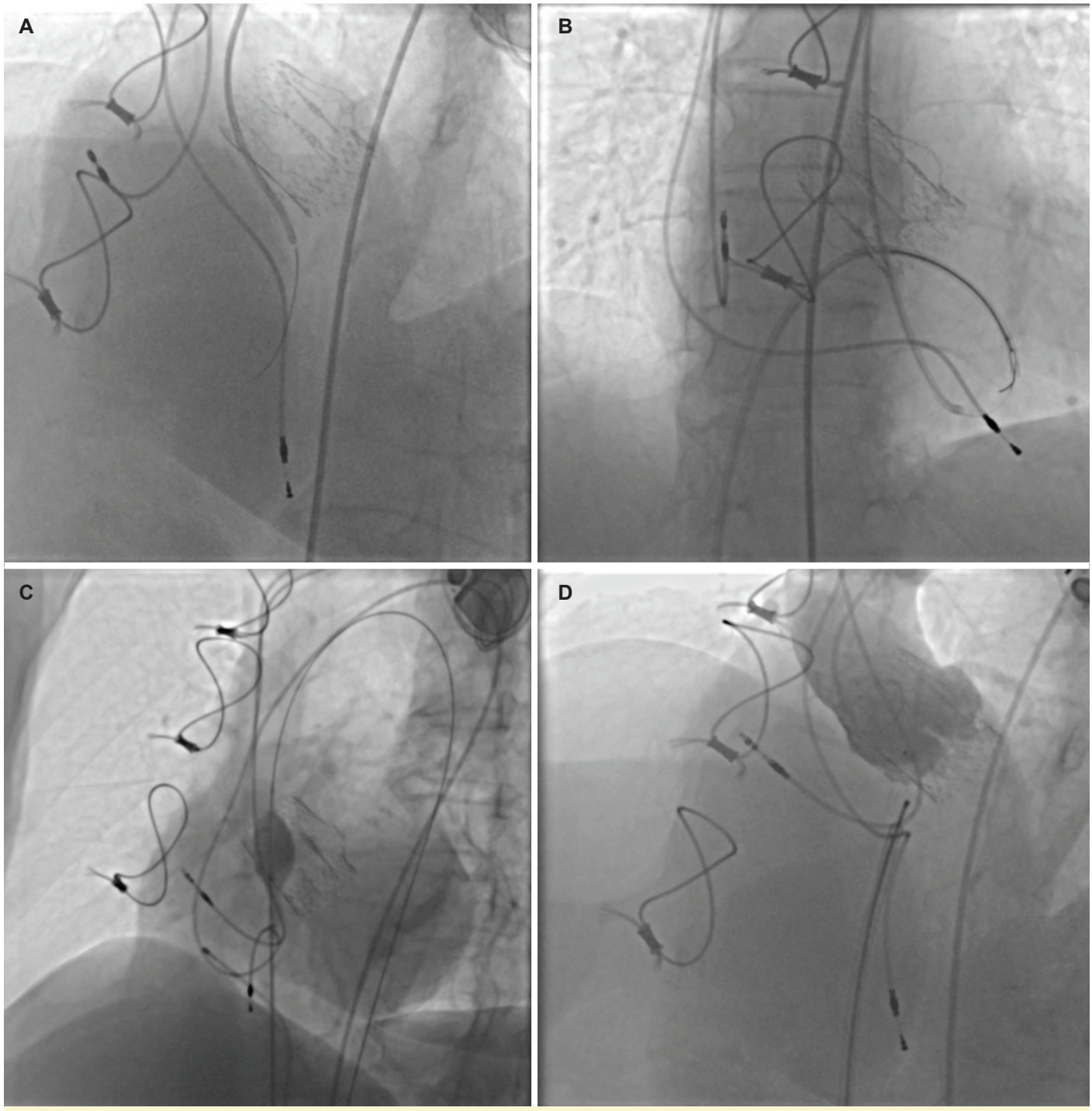

Figure 2. Fluoroscopy image demonstrating the passage of a hydrophilic wire from the defect to the right ventricle (A), Fluoroscopy image capturing the establishment of an arterial-venous wire loop through the ARVF (B), Fluoroscopy image displaying the balloon sizing procedure and confirming the defect's diameter (C), Aortic root angiogram verifying the absence of leakage into the right ventricle (D).

the aortic media and annulus fibrosus.[5] It originates from the right coronary sinus in approximately 70% of cases and from the non-coronary sinus in 25%. If a rupture occurs, a fistula often develops to the RV or atrium.[6] Although surgical repair remains the main treatment option for ruptured SVAs, percutaneous closure serves as an alternative option in patients with a small-sized rupture and no cardiac abnormalities.[7] The use of percutaneous transcatheter closure for ARVFs with various occlusive devices, eliminating the need for open-heart surgery, is becoming increasingly common. A review has indicated that most operators prefer closure devices with a double-disc design. The device should be chosen to be 2-4 mm larger than the

defect, and the closure approach can be either retrograde arterial or transvenous.[8] It is crucial to ensure that there is no significant aortic regurgitation, tricuspid regurgitation, right ventricular outflow tract obstruction, arrhythmia, or coronary compression before releasing the devices. Post-procedural antiplatelet therapy typically includes six months of treatment with ASA and clopidogrel for the first four to six weeks.[8]

Traditionally, the primary imaging modalities for SVA are transthoracic or transesophageal echocardiography. The 'Windsock sign,' a classic finding seen in two-thirds of patients with a rupture, can be identified on transthoracic echocardiography.[9] Ruptures from the right or non-coronary sinus to the right heart chambers can be visualized on anteroposterior or right anterior oblique views with minimal cranial angulation.

When an SVA opens into adjacent cardiac chambers, patients may experience severe hemodynamic changes. Without treatment, ruptured SVAs have a median survival rate of 3.9 years, necessitating early surgical intervention.[10] If left untreated, ruptured SVAs can lead to heart failure and, in about 80% of cases, sudden death.[11]

In the available literature, only one case has been reported involving a patient with a sutureless aortic valve replacement who underwent percutaneous closure.[2] In another instance, percutaneous closure was attempted following sutureless valve implantation in a patient who developed fistulization from the aorta to the right atrium. However, as the procedure was unsuccessful, the patient subsequently underwent surgery.[3] In the only documented case in the literature,[2] the Amplatzer Vascular Plug II device was implanted under the guidance of intracardiac echocardiography (ICE). In our case, we utilized the Amplatzer Duct Occluder and performed balloon sizing to assess the diameter of the defect. In this case, we highlighted that a rupture of the sinus of Valsalva may develop after a sutureless aortic replacement due to extensive leaflet resection. Successful percutaneous fistula closure, guided by fluoroscopic images, is a viable option that can be both time-efficient and cost-effective, eliminating the need for ICE guidance.

Conclusion

The rupture of the SVA and the development of an ARVF, although rare, represent major causes of morbidity and mortality if not promptly diagnosed and treated. While surgery is the primary treatment option, as demonstrated in our case, percutaneous transcatheter closure of an ARVF should also be considered.

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

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