

## Transcatheter closure of ruptured sinus Valsalva aneurysm with retrograde approach

### Yırtılmış Valsalva sinüsü anevrizmasının retrograd yöntemle transkateter kapatılması

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**Summary**– A three-year-old girl with multiple heart malformations admitted to the pediatric cardiology unit because of excessive sweating and fatigue. Abnormal color Doppler flow was detected into the right atrium from the dilated coronary sinus on the echocardiographic examination, and ruptured sinus Valsalva aneurysm (SVA) was diagnosed. Although in most such cases, an antegrade transcatheter approach has been used, a retrograde approach can be used as a cost-effective treatment modality in those cases with selective high-risk surgery. In this report, we present a patient with ruptured SVA, which was closed via Amplatzer vascular plug-4 by retrograde approach.

**S**inus of Valsalva aneurysm (SVA) is thin-walled saccular or tubular outpouchings of the aortic sinus. Open surgical repair with cardiopulmonary bypass is a traditional method for ruptured SVA. Successful transcatheter closure performed with different devices has been increasingly reported as single case reports or in small series. In most cases, an antegrade approach, which requires constructing an arteriovenous femorofemoral wire loop, is used.<sup>[1-4]</sup>

We report herein a three-year-old girl with multiple heart malformations in whom the ruptured SVA was closed using a retrograde approach.

#### CASE REPORT

A three-year-old girl who had been operated for ventricular septal defect, atrial septal defect and double-chambered right ventricle when she was eight months old was presented to the pediatric cardiology unit

**Özet**– Çoklu kalp malformasyonları olan üç yaşında kız hasta, çocuk kardiyolojisi kliniğine aşırı terleme ve çabuk yorulma nedeniyle başvurdu. Transtorasik ekokardiyografide, genişlemiş sol koroner sinüsten sağ atriyuma doğru anormal renkli Doppler akımı belirlendi ve hastaya Valsalva sinüsü anevrizması (VSA) yırtığı tanısı kondu. Bu durumlarda çoğunlukla kateter yoluyla öne doğru yaklaşımla kapama işlemi uygulanmasına rağmen, özellikle cerrahi riski yüksek olan seçilmiş hastalarda geriye doğru yaklaşımla uygulama da etkin ve güvenilir bir tedavi yöntemi olarak kullanılabilir. Bu yazıda, yırtık olmuş VSA'yı Amplatzer vascular plug-4 ile retrograd yaklaşımla kapadığımız hastayı sunduk.

because of excessive sweating and fatigue. The clinical examination revealed tachycardia and tachypnea. S3 heart sound and continuous murmur best heard at the left lower sternal edge were present. A 12-lead electrocardiogram showed sinus rhythm, normal axis and right ventricular hypertrophy. The echocardiographic examination revealed viscerotrial situs solitus, levocardia, dilated and hypertrophic right ventricle, and dilated right coronary sinus with continuous flow into the right atrium, which was shunting left-to-right with a gradient of 52/30 mmHg due to ruptured SVA.

The patient's parents were informed about the treatment options, including surgical repair and percutaneous closure, and it was decided to proceed with a percutaneous approach. The procedure was performed under general anesthesia and transthoracic

#### Abbreviation:

SVA Sinus of Valsalva aneurysm

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echocardiography guidance. Pulmonary vascular resistance was calculated as 1.4 and Qp/Qs as 2.9 in the angiocardiology. The ruptured SVA originating from the right coronary sinus, which fistulized to the right ventricle, was revealed with left ventriculogram and aortogram. The ruptured SVA was tunnel-shaped and measured 14 mm in length and 6.8 mm in width (Video 1\*, Figure 1a).

A 5 Fr right coronary catheter (Cordis; Miami, FL) with a 0.021 hydrophilic wire was advanced into the right atrium through the rupture. Then, the hydrophilic wire was changed with an extra back-up wire. We retrieved the wire and advanced an Amplatzer vascular plug-4 (St. Jude Medical, Inc; St. Paul, MN) distal to the SVA. The defect was closed with an 8 mm in length vascular plug-4 (Video 2\*, Figure 1b). After transthoracic echocardiography confirmed that the device was in the proper position, the device was released. The defect was closed without any complications or aortic regurgitation; however, there was minimal residue shunt of the ruptured SVA in the follow-up aortography and transthoracic echocardiography. Total fluoroscopy time was 7.3 minutes. She was discharged with oral aspirin prophylaxis for six months. Trivial residue shunt in the echocardiography

was seen in the follow-up. Trace aortic regurgitation and moderate tricuspid regurgitation recovered after the procedure. Her symptoms improved and she had no recurrence or other complications during the two-year follow-up period.

## DISCUSSION

Sinus of Valsalva aneurysm is usually a congenital anomaly and accounts for about 1% (range, 0.1-3.5%) of congenital anomalies of the heart. The SVA generally ruptures into the right side of the heart and results in left-to-right shunt. When the SVA ruptures, nearly 80% of the patients become symptomatic because of important hemodynamic effects. The ruptured SVA (95%) mostly arises from the right coronary sinus.<sup>[4,5]</sup> With the recent advances in percutaneous closure of other left-to-right shunts, it is expected that percutaneous closure might be a good choice for the treatment of this rare anomaly.

The occurrence of a congenital ruptured SVA is related to incomplete fusion of the distal bulbar septum (primitive bulbous cordis), with truncal ridges or lack of fusion of the tunica media and annulus fibrosis of the aortic valve.<sup>[5,6]</sup> Also, in our case, it was thought that the SVA originating from the right coronary si-



**Figure 1.** (A) Left ventriculogram views depicting the ruptured sinus Valsalva, which was tunnel-shaped, with regurgitation into the right atrium. (B) Aortography view showing closure of the ruptured sinus Valsalva with slight residual shunt.

nus ruptured to the right ventricle dissecting the fibrous tissue in the atrioventricular sulcus. Ruptured SVA can be associated with various congenital heart defects. We are not aware of any other ruptured SVA case associated with double-chambered right ventricle or any other multiple cardiac defects in the related literature.

The clinical presentation of ruptured SVA is varied and can be catastrophic with important left heart failure outcome. Our patient had the typical presentation of ruptured SVA, i.e., excessive sweating, palpitation and fatigue with symptoms of left heart failure. Since 1956, the main treatment option has been surgical repair using prosthetic or pericardial patches with cardiopulmonary bypass.<sup>[5-7]</sup> However, surgical treatment carries morbidities of both sternotomy and cardiopulmonary bypass. The transcatheter procedure, in addition to its diagnostic confirmation and assessment of shunt and pulmonary vascular bed, allows closure of the defect in suitable cases. An antegrade approach is used in most of the cases.<sup>[1-3]</sup> This method necessitates the use of an additional snare and arteriovenous loop and prolongs fluoroscopy time. The retrograde approach can be safe in appropriate cases like ours.<sup>[8-10]</sup> In the literature, depending on the anatomy of the lesion, use of different devices, such as Raskind umbrella, Gianturco coil, Amplatzer duct occluder, muscular ventricular septal occluder, Amplatzer septal occluder, and patent foramen ovale occluder, has been reported for closure of the ruptured SVA.<sup>[1-3, 8-10]</sup> We preferred to use vascular plug-4 for closure of the ruptured SVA because it has a low metallic load and is safer.

Although no complications were encountered, several complications associated with the percutaneous procedure can be seen, such as failure of the device to deploy, residual shunting, procedure-related aortic regurgitation, coronary encroachment by the device, dysrhythmia, and bleeding.<sup>[5-10]</sup> In conclusion, transcatheter closure using a retrograde approach is a cost-effective modality for ruptured SVA.

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**\*Supplementary video file associated with this article can be found in the online version of the journal.**

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**Anahtar sözcükler:** Aort anevrizması; aort yırtığı/cerrahi; kalp septal defekti; Valsalva sinüsü; ventriküler/tedavi.