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Successful transcatheter closure of very large ruptured sinus of Valsalva aneurysm using the Cocoon Duct Occluder (CDO)

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Abstract

Sinus of Valsalva aneurysm, usually a congenital anomaly, almost always ruptures into the right side of the heart causing a left-to-right shunt with profound hemodynamic consequences. With the availability of devices and hardware, transcatheter closure is gradually replacing surgical one. Till now, most of closures have been performed by Amplatzer duct occluder. To the best of our knowledge, the present case is first to be reported with this rare defect undergoing successful transcatheter closure of largest ruptured sinus of Valsalva aneurysm arising from right coronary sinus by using 20/18 mm Cocoon Duct Occluder (Vascular Innovations, Nonthaburi, Thailand).

Key words: sinus of Valsalva aneurysm, Amplatzer duct occluder, Cocoon Duct Occluder, transcatheter interruption

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Introduction

Sinus of Valsalva aneurysm, a blind pouch projecting from one of the sinuses into the right atrium or right ventricle is an uncommon congenital heart defect (CHD) accounting for < 1% of all CHDs. It forms due to congenital weakness at the junction of the aortic media and annulus fibrosus [1]. Aneurysms mostly arises from right coronary sinus (RCS) in 70%, non-coronary sinus (NCS) in 29%, and rarely from left coronary sinus in 1% [2]. Ruptured sinus of Valsalva aneurysm is a rare cardiac anomaly which may open into the right ventricle or right atrium, whereas aneurysms of non-coronary sinus (NCS) rupture into the right atrium and rarely into the pulmonary artery, left ventricle, left atrium, or pericardial cavity [3]. Presentation varies from asymptomatic findings, sudden onset of chest pain to acute heart failure, which can rapidly worsen to cause death which usually occurs within 1 year if left untreated [3]. Though surgery is the gold standard, percutaneous transcatheter closure (TCC) has now become equally efficacious with fewer complications. Most of the data on TCC of RSOVA is by Amplatzer duct occluder (ADO).

Case study

A 24-year-old woman with no other significant co morbidities presented with history of palpitations and breathlessness on exertion of NYHA class III for 4 month duration. On examination she was found to have continuous murmur in the right second, third and fourth intercostal space with peripheral signs of aortic run-off. Electrocardiogram showed left ventricular hypertrophy with right atrial enlargement and chest X-ray showed cardiomegaly with right atrial enlargement. 2D TTE showed dilated right atrium and

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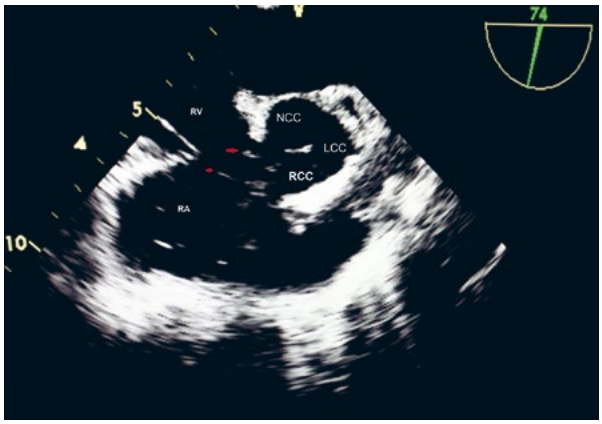

Figure 1. TEE showing aneurysmally dilated right sinus of Valsalva communicating into right atrium with a classical *windsock deformity* (red arrow)

right ventricle and continuous shunt opening into right atrium with peak systolic gradient of 68 mm Hg and end diastolic gradient of 12 mm Hg. Transesophageal echocardiography (TEE) showed aneurysmally dilated right sinus of Valsalva which was communicating into right atrium with continuous flow revealing a classical *windsock deformity* (Fig. 1). Subsequently she underwent aortic root angiography which confirmed aneurysm of right coronary sinus rupturing into right atrium forming a tunnel. Right coronary artery had its origin just above the aneurysm (Figs. 2, 3). Aortogram was performed in left anterior oblique (LAO) with cranial tilt projection and right anterior oblique (RAO) view to profile the communication. RSOV defect was measured by QCA (quantitative coronary analysis) and TEE to be 16–18 mm. Device closure was planned with informed consent as there were no associated defects requiring surgical correction (like ventricular septal defect [VSD] or significant aortic regurgitation [AR]). The femoral vein and artery were accessed with 8 and 5F sheaths respectively. Intravenous heparin (100 IU/kg) and ceftriaxone were given. The defect was crossed from the aortic side using a 5F Judkins right (JR) coronary catheter over a 0.035" straight tip glide wire (Terumo Inc, Japan). With catheter across RSOV, exchange length wire (330 cm, 0.035" Terumo noodle wire) was passed into distal inferior vena cava (IVC) and snared with 2 cm Amplatz gooseneck snare (Microvena, MN, USA) and brought out of right femoral vein sheath establishing a stable arteriovenous wire loop. 12F long Cocoon delivery sheath (Vasc Innovation, Thailand) was introduced over the exchange length wire from the venous side into ascending aorta across the defect. The size of Cocoon Duct Occluder (CDO; Vasc Innovation, Thailand) selected was 20/18 mm so that its aortic segment was 2 mm larger than diameter of the defect. CDO with its

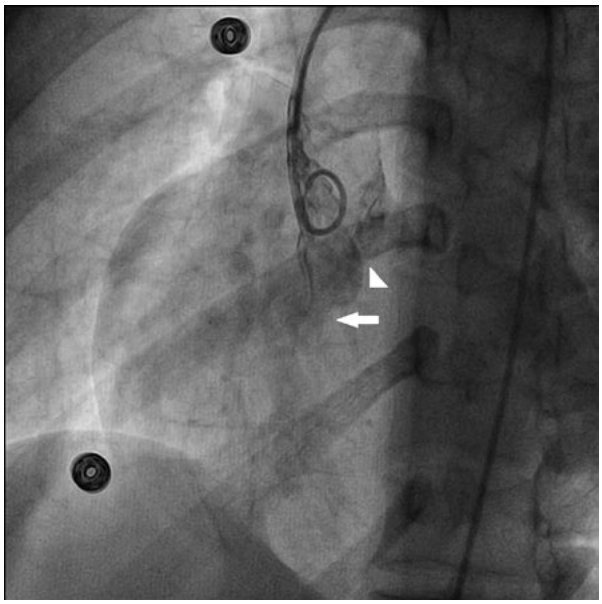

Figure 2. Aortic root angiogram showing aneurysmal sac (arrowhead) of right coronary sinus rupturing into right atrium forming a tunnel (arrow). Right coronary artery had its origin just above the aneurysm

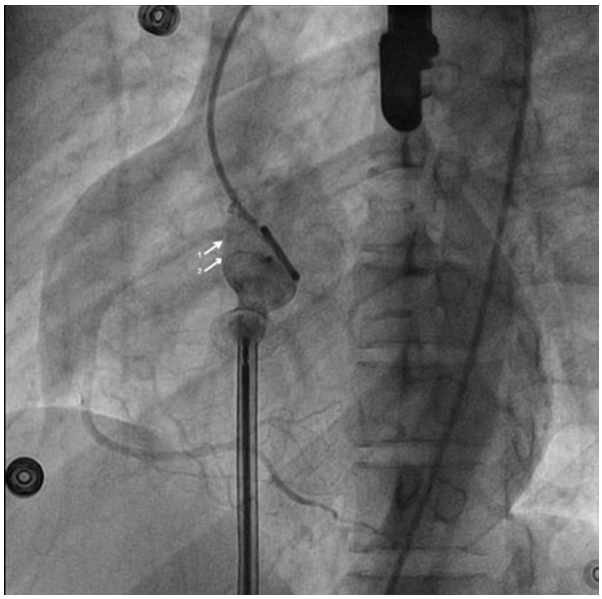

Figure 3. Aortic root angiogram showing origin of right coronary artery (1) just above the aneurysm (2)

attached delivery cable was then inserted through the delivery sheath, and its aortic disk was deployed in the ascending aorta. The whole assembly was pulled back and positioned carefully till the aortic disk blocked the aortic end of the SOVA and it was ensured that aortic valve leaflets were free as seen on TEE (Fig. 4). After confirming the precise placement, the rest of the ADO was deployed on the right side across the defect. During this maneuver, a gentle traction was exerted on the

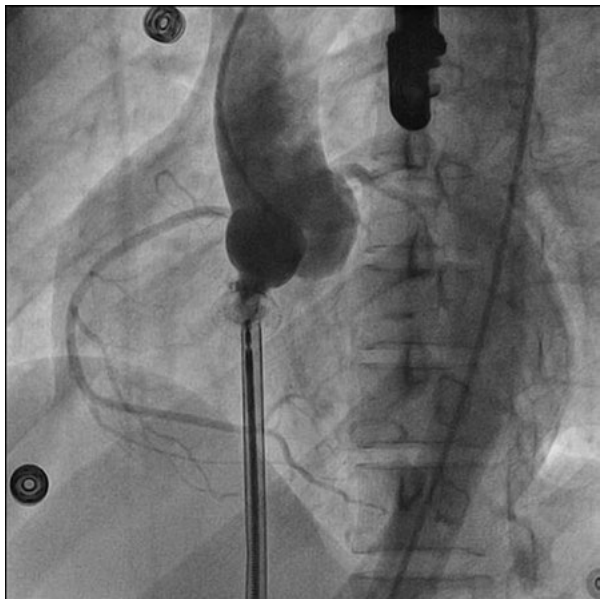

Figure 4. Precisely placed CDO across the defect

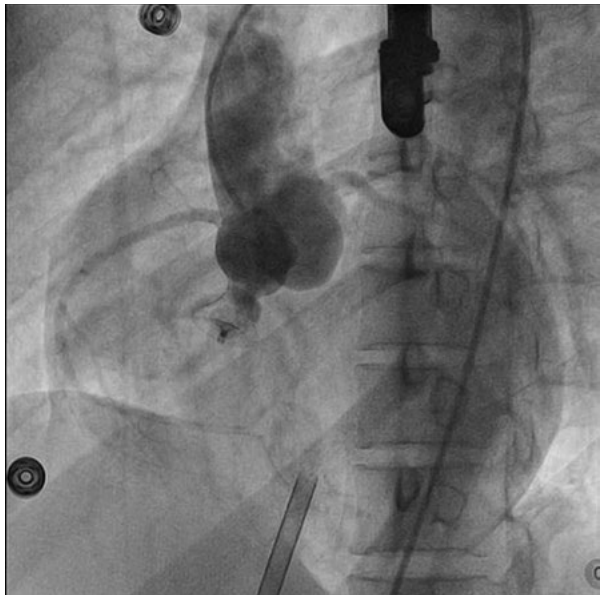

Figure 5. Properly deployed device with mild flow on root angiogram

delivery cable, but taking special care to ensure seating of the aortic disk on the aortic side without slippage into the aneurysm. After 10-min delay, aortic root angiogram was again performed to ensure proper device position and make certain that there was no significant AR, tricuspid regurgitation (TR), or any encroachment on right coronary artery which was also confirmed on TEE. The CDO was then released from the delivery cable by turning the cable counter clockwise using the pin vise (Fig. 5). There was mild flow across the defect which subsequent sealed as echo next day showed no

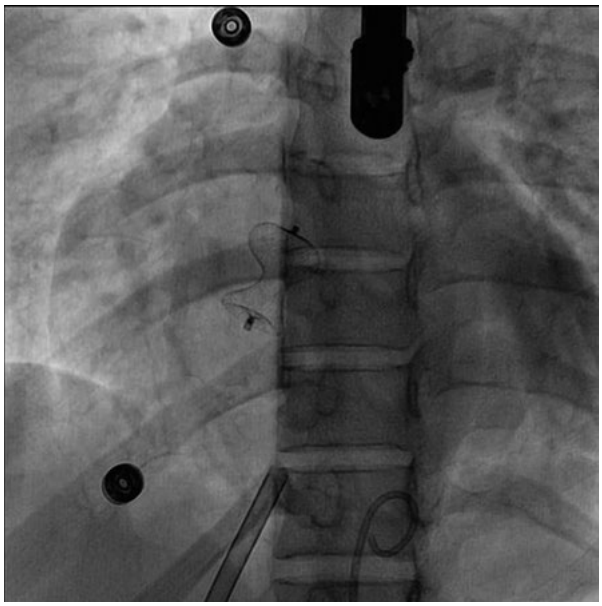

Figure 6. Properly deployed device

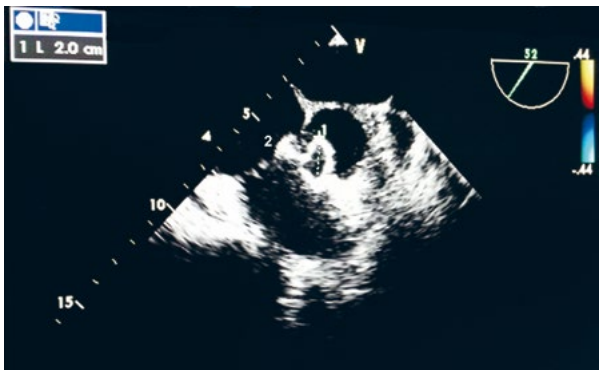

Figure 7. Properly deployed device on TEE

residual flow across the device (Figs. 6, 7). Patient was discharged in stable position on next day with advice of aspirin 75 mg once daily for 6 months.

Discussion

RSOV is more common in males with mean age of rupture being third decade [4]. SVA is more often congenital than acquired in origin. Acquired causes include infective endocarditis, syphilis, trauma, connective tissue disorders, Marfan syndrome, atherosclerosis and cystic medial necrosis. SVA comes to clinical attention because of asymptomatic or symptomatic unruptured aneurysm. Patients succumb to death from congestive heart failure usually within a year after rupture; however, longer survival after small slow perforation is well described. SVA can present in various ways such as rupture into cardiac chamber with subsequent cardiac failure, acute coronary syndrome, cardiac tamponade, heart block,

aortic regurgitation and right ventricular outflow tract obstruction [5]. Majority of SVA arising from right coronary sinus ruptures in right ventricle and those arising from non-coronary sinus ruptures into RA [6].

The technique is similar to TCC of perimembranous VSD, although the defect is located just above the aortic valve instead of below. Periprocedural TEE and color Doppler interrogation helps us in sizing the defect, device selection (2–4 mm larger than the aortic end), delineating the SOVA anatomy in regards to its neighboring structures namely the aortic valve, tricuspid valve, and RVOT, ensuring proper seating of the aortic disk on the aortic side without slipping into the body of the aneurysm, and most importantly, monitoring AR and TR occurrence and residual shunting on color Doppler. Although Arora et al. [7] had used TTE, we believe that TEE guidance makes procedure much comfortable for the operators. Unlike Arora et al. [7] we did not utilize balloon sizing of the defect since we were able to size the defect well on TEE. We attempted to close the ruptured SOVA at the aortic end similar to a 'surgeon's repair' since closure at the rupture site (exit point) would leave behind an aneurismal sac exposed to arterial pressure with a potential to rupture at another site in the long term [8].

The CDO is a surgical permanent implant. It is a platinum coated self-expandable, mushroom shaped device made from a Nitinol wire mesh. A mushroom shaped retention skirt assures secure positioning at the aortic orifice of patent ductus arteriosus. The communication is closed by the introduction of thrombosis which is accomplished by multiple poly propylene patches sewn securely to the side of the device. Nano Platinum coating provides superior biocompatible properties compared to bare nitinol and prevents nickel leaching into the blood stream and corrosion of nitinol wire frame in long term implants. Platinum provides better radio-opacity which enables easy positioning of the device in the defect. TCC of ruptured sinus of Valsalva can be safely and effectively done using CDO and a good alternative to surgery. After extensive search in literature and to the best of our knowledge, this is the largest report of RSOVA closure using CDO.

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