Transcatheter Closure of Ruptured Sinus of Valsalva Aneurysm

Report of two cases

*Khalfan S. Al-Senaidi,1 Abdullah Al-Farqani,2 Madan Maddali,1 Salim Al-Maskary2

ABSTRACT: A ruptured sinus of Valsalva aneurysm (RSVA) is a rare cardiac anomaly. Traditionally, RSVAs were repaired surgically; however, percutaneous transcatheter closure is the current treatment of choice. We report two cases of RSVA which were closed using this approach. The first case was a 45-year-old female who presented to the Royal Hospital, Muscat, Oman, in 2014 with a RSVA in the right ventricle. The second case was a 39-year-old male who was admitted to the Sultan Qaboos University Hospital, Muscat, in 2015 with a large multifenestrated RSVA extending into the right ventricle outflow tract. Each patient underwent transcatheter cardiac catheterisation using three-dimensional echocardiography. Both interventions were technically successful; however, the second patient required a subsequent surgery due to the continuing presence of a significant shunt. Transcatheter closure of RSVAs is an effective alternative to surgical repair, although large multifenestrated RSVAs should be repaired surgically to ensure complete closure.

Keywords: Sinus of Valsalva; Ruptured Aneurysm; Cardiac Catheterization; Three-Dimensional Echocardiography; Case Report; Oman.

First described in 1839, a ruptured sinus of Valsalva aneurysm (RSVA) is a rare but well-known clinical condition, with the rupture usually occurring in the right-sided cardiac chamber. These types of aneurysms are thin-walled sacular or tubular outpouchings of the aortic sinuses and can be either congenital or acquired.2 Echo-cardiography can accurately identify these lesions and confirm a diagnosis of RSVA. Cardiac surgical repair is the conventional method of treatment; however, transcatheter closure has more recently been deemed the optimal choice of treatment.3 This report presents two cases of RSVA in which transcatheter closure was used. Complete closure of the defect was achieved in the first case. For the second case, which involved a patient with a multifenestrated aneurysmal defect, two devices were used during catheterisation. However, complete closure could not be achieved.

Case 1

A 45 year-old female with no known medical conditions presented to the Emergency Department of the Royal Hospital, Muscat, Oman, in 2014 with a one-day history of chest pain and shortness of breath. There was no history of fever. Her blood pressure was 120/55 mmHg and her heart rate was 85 beats per minute. A clinical examination revealed normal heart sounds, although a continuous murmur was audible over the third left intercostal space. An electrocardiogram and chest X-ray were normal with no features suggestive of a myocardial infarction. Her
Troponin T levels were within the expected range (<5 pg/mL). Transthoracic echocardiography (TTE) and transoesophageal echocardiography (TOE) revealed a RSVA extending from the right sinus into the right ventricle under the septal leaflet of the tricuspid valve. The aneurysm had a windsock appearance, with the aortic end measuring 10 mm and the opening into the right ventricle measuring 6 mm [Figure 1A]. She had a trileaflet aortic valve with no regurgitation. No ventricular septal defects, pericardial effusions or intracardiac vegetations were seen. The right and left ventricles were not dilated and demonstrated normal systolic function.

After discussion with cardiothoracic surgeons, a transcutaneous cardiac catheterisation intervention was performed. The patient was put under general anaesthesia, with the right femoral artery and vein punctured and a 6F short sheath secured. Unfractionated heparin and cefradine were administered intravenously at doses of 100 IU/kg and 1,000 mg, respectively. Two- and three-dimensional TOE scans confirmed the previous echocardiographic findings and haemodynamic data showed an increased left ventricular end-diastolic pressure of 15 mmHg and a pulmonary to systemic blood flow (Qp:Qs) ratio of 3:1. An aortic root angiogram clearly delineated the RSVA, which was located far away from the right coronary artery with similar measurements to those observed via TOE. The RSVA was closed using a size 12/10 Cocoon Duct Occluder (Aeon World Group Corp., Taguig City, Manila, Philippines), which was inserted antegrade from the venous side. The RSVA was crossed from the aortic side with a 5F Cordis™ Judkins Right JR4 catheter (Cordis Corp., California, USA) using 0.89 mm x 260 cm of guidewire (Terumo Interventional Systems, Terumo Medical Corp., Tokyo, Japan). An arteriovenous loop was established after the wire was snared from the left pulmonary artery. A 7F Mullins sheath (Cook Group, Bloomington, Indiana, USA) was placed into the ascending aorta before the occluder was introduced; the skirt of the device was opened in the ascending aorta under fluoroscopic and TOE guidance. The device was pulled as one unit into the RSVA and the sheath retracted over the cable to deploy the rest of the device. Three-dimensional TOE and an angiogram confirmed that the device was in a satisfactory position, with minimal flow through the device and no aortic valve regurgitation [Figures 1B and C].

The following day, TTE confirmed that the device was well-positioned, with total occlusion of the RSVA and no aortic or tricuspid valve regurgitation. The patient was discharged and prescribed a six-month course of aspirin (5 mg/kg per day) and a two-month course of clopidogrel (75 mg per day), with close follow-up.

Case 2

A 39-year-old man was admitted to the Sultan Qaboos University Hospital, Muscat, in 2015 with a two-month history of chest pain, shortness of breath and exercise intolerance. Clinically, he showed signs of aortic regurgitation, with an early diastolic murmur along the left sternal border. Using TTE, a RSVA extending into the right ventricular outflow tract (RVOT) was observed, with a dilated left ventricle. A mitral annulus (5 cm) and mild mitral and aortic valve regurgitation were also noted. The aneurysm had two large fenestrations of 8 mm in diameter each, in addition to multiple smaller defects. The mouth of the main aneurysm was 17 mm in diameter and the right coronary artery was in close proximity to the opening of the aneurysm. The aneurysm protruded into the RVOT but did not obstruct it.

The patient underwent transcutaneous cardiac catheterisation of the ruptured aneurysm under TOE guidance with on-site surgical back-up. A TOE scan...
confirmed the TTE findings and showed that the RSVA was 3.4 cm with a 17 mm mouth [Figure 2A]. The right femoral artery and vein as well as the left femoral artery were accessed using 8F, 10F and 6F sheaths, respectively. The coronary arteries were normal and the Qp:Qs ratio was 5.2:1, with a pulmonary vascular resistance index of 1.6 Wood units. As previously described for the first patient, the fenestrated defect was crossed and an arteriovenous loop established. The first fenestration was closed with a 23/25 mm Occlutech® Figulla® PFO Occluder (Occlutech GmbH, Jena, Germany), inserted antegradely from the venous side. A large shunt in the second fenestration was identified via TOE, necessitating the use of a second device; hence, an 18mm AMPLATZER™ PFO Occluder (St. Jude Medical Inc., St. Paul, Minnesota, USA) was deployed retrogradely from the arterial side. This device was deployed retrogradely in order to close the aneurysm from the RVOT towards the left ventricle and to secure the two devices in a more stable position. The right coronary artery was not affected by the devices and both devices were placed in suitable positions [Figures 2B and C]. Following the intervention, the Qp:Qs ratio decreased to 2.2:1 and TEE revealed mild aortic regurgitation.

The patient was discharged the following day, after a TTE scan confirmed the position of the devices. However, a mild-to-moderate shunt was present across the aneurysm, with no RVOT obstruction. The mild aortic regurgitation persisted. The patient was prescribed 75 mg of aspirin daily for six months and 75 mg of clopidogrel daily for two months. On follow-up, the patient reported significant improvement in his symptoms. Echocardiography confirmed that the two devices were still in a satisfactory position across the aneurysm with no RVOT obstruction; however, there was a significant shunt across the other small defects. The patient subsequently underwent successful surgical correction of the aneurysm a few months later.

### Discussion

Congenital RSVAS account for 0.1–3.5% of congenital heart defects and have a higher incidence in Asian versus Western populations. These aneurysms usually rupture into the right side of the heart, producing a left-to-right shunt which has a profound haemodynamic effect. Drainage sites can be single or multiple, with the right ventricle being the most common. Previous research has described a wide spectrum of presentations, ranging from an asymptomatic murmur to cardiogenic shock or even sudden cardiac death. Lillehei et al. reported the first successful repair of RSVAS for three patients in 1957. Traditionally, surgical closure has been the mainstay of treatment for RSVAS, with an operative mortality rate of <5% and excellent long-term outcomes. Nevertheless, these patients remain at risk of prolonged hospital stays and postoperative complications such as chest pain and septicemia, making percutaneous device closure an attractive alternative. The transcatheter closure of a RSA was first reported by Cullen et al. in 1994 using a Rashkind umbrella device. Advances in cardiac catheterisation technology have resulted in a wide range of devices and coils which can be used to close a RSA. Retrograde and combined minimally invasive surgery hybrid approaches have been described in certain cases.
jet in order to facilitate the selection of occluders for percutaneous closure.\textsuperscript{19} The four types of shunt jets identified are: type I, window-like; type II, aneurysmal; type III, tubular; and type IV, other rare conditions.\textsuperscript{19} With regards to the current cases, the first patient had a type III shunt jet and the second patient had a type II shunt jet.

The current report describes two patients with RSVAs in which a transcatheter approach was used to close the aneurysms. In the first case, the RSVG was closed using an occluder made of platinum-coated nitinol wires. This is a self-expanding double-disk device connected by a joint at the centre of the disks, which are filled with polypropylene fabric to increase thrombogenicity. Complete closure of the defect was achieved in this case. The second patient had a complex multifenestrated RSVG with two large defects and additional smaller defects. The RSVG penetrated into the RVOT and required two devices to close the larger defects. Initially, the interventionist intended to close the aneurysm at its entry point; however, due to the close proximity of the right coronary ostia to the mouth of the aneurysm, it was decided to close the RSVG at the exit point instead. Patent foramen ovale occluders were used as they have large retention discs on both sides which are nearly equal in size. As the aneurysm was multifenestrated, the devices were positioned in order to cover as many of the smaller defects as possible. Nevertheless, the patient continued to experience a significant shunt through the small defects and was referred for further surgical correction. These two cases demonstrate the two extremes of RSVG in terms of complexity. As observed with the second patient, large multifenestrated RSVAs should be closed surgically—if closure at the point of entry is not possible—in order to prevent the occurrence of smaller defects. In most cases of RSVG, the coronary ostia are quite high and device impingement occurs rarely. Ventricular septal defects often coexist with RSVAs, especially in patients with significant aortic valve regurgitation.\textsuperscript{20} Thankfully, neither patient in the current report had a ventricular septal defect.

**Conclusion**

Transcatheter closure is a safe and effective treatment option for patients with RSVAs. Real-time three-dimensional echocardiography allows easy visualisation of the defect and is helpful in guiding deployment of the device. However, surgical repair is recommended for large multifenestrated defects to prevent the development of further small defects after closure. The long-term follow-up of RSVG patients is mandatory to ensure complete closure of the aneurysm.

**References**


