

Successful Percutaneous Transcatheter Device Closure of Ruptured Sinus of Valsalva Aneurysm

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Abstract: Ruptured Sinus Of Valsalva (RSOV) is a rare but clinically well recognized entity which predominantly affects adolescent males caused by congenital weakness in the aortic media leading to progressive dilatation of sinus eventually leading to rupture in to right sided cardiac chambers of the heart resulting in a left to right shunt with grave hemodynamic compromise. It usually requires surgical correction but transcatheter device closure is an alternative option. Here we present a case of 52 year female who presented to cardiac ICU with shortness of breath and palpitations. She was diagnosed as a case of ruptured sinus of valsalva aneurysm (SVA) draining from non coronary sinus into right atrium. She underwent successful percutaneous device closure with 14x16 mm lifetech PDA occluder and complete closure of the defect was achieved with no residual shunt, device embolization or infective endocarditis and no progression of AR at follow-up. Transcatheter closure of ruptured SVA is a safe and effective alternative to surgical repair in appropriately selected cases and long-term follow-up is mandatory to ensure complete closure of the aneurysm.

Keywords: Sinus of Valsalva aneurysm (SVA), Transcatheter closure (TCC), Rupture, Echocardiography, Cardiac catheterization, Device.

INTRODUCTION

Ruptured Sinus Of Valsalva (RSOV) is a rare but clinically well recognized entity which predominantly affects adolescent males with incidence of 0.3-0.7/1000 general population. It is caused by congenital weakness in the aortic media leading to progressive dilatation of sinus eventually leading to rupture.

A congenital Sinus of valsalva aneurysm (SVA) is usually clinically silent but may vary from asymptomatic dilation detected in routine two-dimensional (2D) echocardiography, to symptomatic presentations related to the compression of adjacent structures or rupture in to right sided cardiac chambers of the heart resulting in a left to right shunt with grave hemodynamic compromise.

2D Echocardiography with Colour Doppler is used to establish the diagnosis and quantify the defect dimensions. Open surgical repair with cardiopulmonary bypass has been conventionally the mainstay of therapy for RSOV. However, successful transcatheter closure (TCC) is being increasingly reported in recent times [1-6].

CASE REPORT

A 52 year old female patient presented with complaints of dyspnea on exertion and palpitations since last 6 months which initially was NYHA Class I and gradually progressed to NYHA class III at the time of presentation. A Clinical Examination revealed her heart rate was 136/min regular, respiratory rate of 26/min, BP 160/60 with Grade IV/VI harsh continuous murmur best heard in left lower parasternal area. Chest X ray showed cardiomegaly, ECG showed sinus tachycardia and LVH with strain. Transthoracic echocardiography (TTE) showed Situs solitus, atrioventricular and ventriculoarterial concordance with left sided aortic arch, aortic root was dilated (4.1cm) with Grade II AR. A fistulous tract was seen between non coronary sinus (aortic end-10 mm) and right atrium (atrial end-6 mm). Left ventricle was dilated with good biventricular function, Grade I mitral regurgitation, Pulmonary artery was dilated (2.8cm) with Mild Pulmonary arterial hypertension (AcT-76 msec). Colour Doppler revealed high velocity jet across the defect from aortic root to right atrium (Figure 1).

Cardiac catheterization showed normal coronaries with QP/QS ratio of 3:1. An aortic root angiogram in left anterior oblique with cranial tilt

projection showed ROSV draining from noncoronary sinus into right atrium.



Fig-1: TTE, parasternal short-axis view, displaying an RSOV (arrow), with colour Doppler demonstrating shunting into right atrium near the tricuspid valve

Procedure

The procedure was performed under local anaesthesia. The right femoral vein and left femoral artery were accessed percutaneously with 7 F and 6 F introducer sheaths, respectively. Intravenous heparin 100 IU/kg and antibiotic were given. An aortic root angiogram was taken with 6 F pigtail catheter in left anterior oblique with cranial tilt projection (Figure 2A) which showed draining from noncoronary sinus in to right atrium. The size was assessed using both Trans-thoracic echo (TTE) and angiography for device selection with aortic end diameter larger than 2-4 mm larger than the size measured.

The defect was crossed from the aortic side using a 5 F Amplatz coronary catheter and a 0.035 × 260 cm J tip guide wire (Terumo Inc., Japan) and was passed into superior vena cava (SVC). Snaring was done with Goose neck snare from SVC (Figure 2B) and

exteriorized out from the femoral vein. An arteriovenous wire loop was established (Figure 2C). Over the Terumo wire, 5 F Multipurpose catheter was passed and crossed the defect and kept in ascending aorta. Now Terumo wire was exchanged with 0.035 x 260 cm Amplatz Super Stiff wire.

Over the Amplatz super stiff wire, through 9F PDA device delivery sheath (Figure 2D), PDA device 14 x16 mm was crossed across the defect. The sheath was then gradually withdrawn resulting in opening of the device at the aortic end followed by the atrial end (Figure 2E). With the device still attached to its delivery system, an aortic root angiogram was done which showed no residual shunt and no impingement of the coronary ostia (Figure 2F). TTE showed no impedance of flow across the tricuspid valve or interference with the motion of the tricuspid valve leaflet by the device (Figure 3).

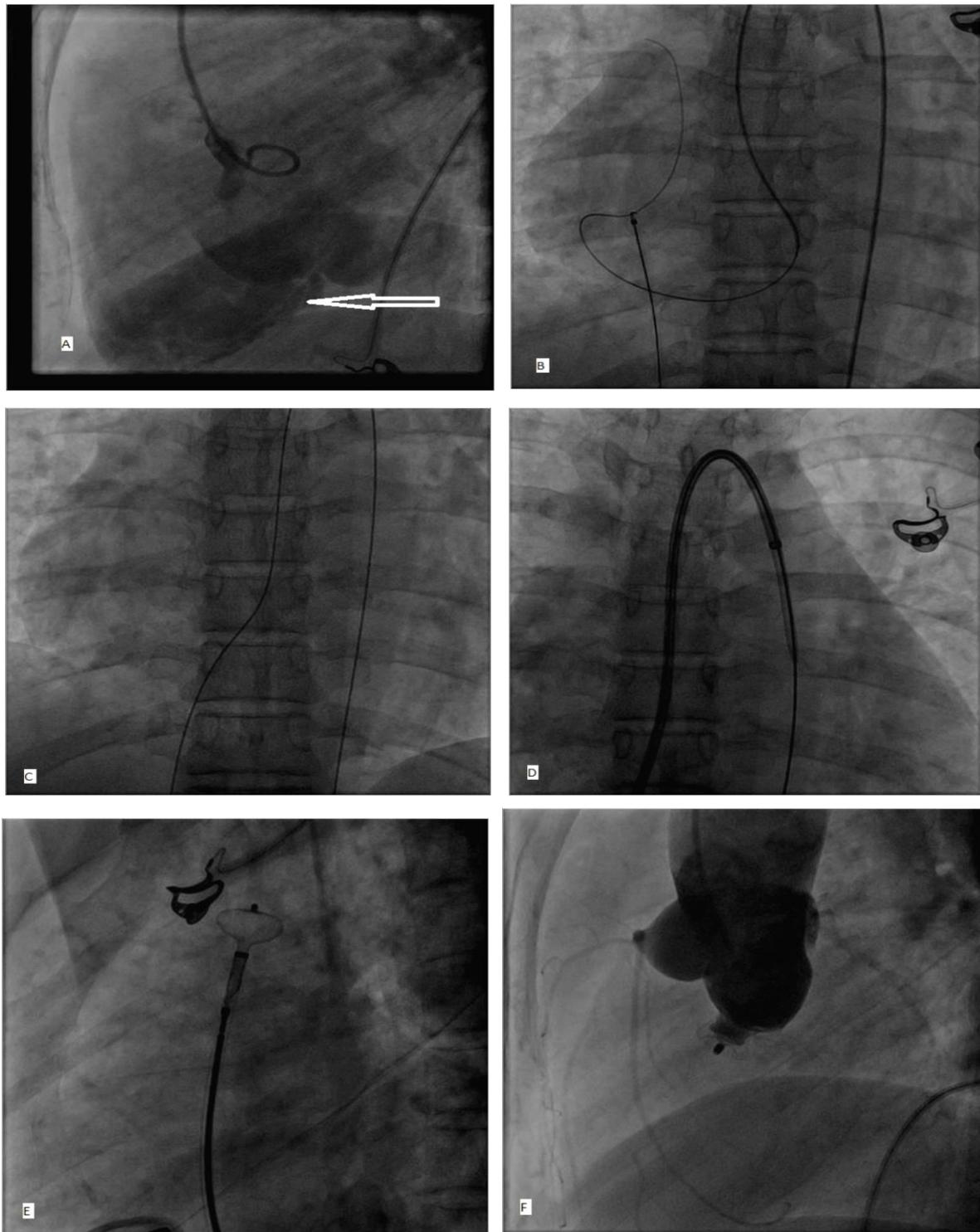


Fig-2: (A) Aortic root angio in LAO view showing noncoronary sinus rupturing into the RA, (B) snaring from SVC, (C) arteriovenous wire loop, (D) PDA device delivery sheath, (E) Device positioning across RSOV and (F) Aortic root angio after device deployment showing no residual shunt across RSOV.

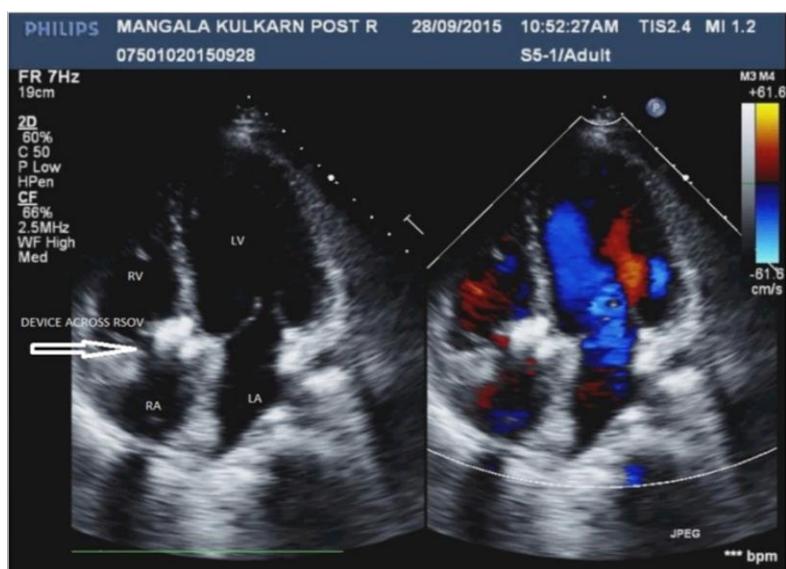


Fig-3: TTE, apical 4 chamber view, showing the device in situ (arrow) across the RSOV with colour doppler showing no residual shunt.

Following this, the device was delivered. Post Procedure course was uneventful and repeat echocardiography showed the device in situ with no residual shunt. The patient was discharged after 48 hours with Tab Aspirin 150 mg OD for 6 months.

Follow-up

Patient was followed up at 1, 3, 6 months and at one year. On follow-up, the patient reported significant improvement in symptoms. She is stable and in NYHA class I and Echocardiography showed no residual shunt, device embolization or infective endocarditis and no progression of AR. Patient was advised to follow-up annually.

DISCUSSION

The incidence of ruptured SVA is five times higher in Asian countries (0.46-3.5% in Eastern and 0.14-0.23% in Western areas) [7, 8] with male/female ratio 4:1 [9]. SVA affect the right sinus or noncoronary

sinus in 90-95% of cases and the left sinus in < 5%. They are associated with other heart defects, VSD (30-60%), aortic valve abnormalities such as aortic regurgitation (20-30%), bicuspid valve (10%), aortic stenosis (6.5%) [9]. They may also be associated with pulmonary stenosis (9.7%), coarctation of the aorta (6.5%), persistence of the ductus arteriosus (3.2%), tricuspid regurgitation (3.2%) and interatrial defect [9]. Rupture of a SVA occurs principally at the RV (60%) or at the RA (29%), the LA (6%), and LV (4%) or at the pericardium (1%) [9].

The classification of SVA has been proposed by Sakakibara and Konno in 1962, describing four types according to the coronary sinus affected and the area where they protrude or rupture (Table -1) which were modified by Xin-Jin L, Xuan *et al.* in 2013 [10]. TTE and transesophageal echocardiogram (TEE) have a diagnostic accuracy of 75% and 90%, respectively [9].

Table 1 Classification systems for RSVA

Type	Sakakibara classification	Modified Sakakibara classification
I	Originating from left part of right coronary sinus; protruding into conus of right ventricle, just beneath commissure of right and left pulmonary valves	Protrusion and rupture into right ventricle just beneath pulmonary valve
II	Originating from central part of right coronary sinus; protruding into right ventricle; penetrating crista supraventricularis	Penetration and rupture into or just beneath crista supraventricularis of right ventricle
IIIv	Originating from posterior part of right coronary sinus; protruding into right ventricle, just beneath septal leaflet of tricuspid valve; penetrating membranous septum	Penetration and rupture into right ventricle adjacent to or at tricuspid annulus
IIIa	Originating from posterior part of right coronary sinus; protruding into right atrium, near commissure of septal and anterior leaflets of tricuspid valve	Penetration and rupture into right atrium adjacent to or at tricuspid annulus
IV	Originating from right part of noncoronary sinus; protruding into right atrium, near septal leaflets of tricuspid valve	Protrusion and rupture into right atrium
V		Other rare conditions (eg, rupture into left atrium, pulmonary artery, left ventricle, or other structures)

Traditionally, open surgical closure with cardiopulmonary bypass has been the mainstay of treatment for RSVAs with an operative mortality rate of <5% and excellent long-term outcomes [7, 11]. Nevertheless, these patients remain at risk of prolonged hospital stays and postoperative complications such as chest pain and septicaemia, making percutaneous device closure an attractive alternative [7]. The transcatheter closure of a RSVA was first reported by Cullen *et al.* in 1994 using a Rashkind umbrella device¹. Advances in cardiac catheterization technology have resulted in a wide range of devices and coils which can be used to close a RSVA [3, 12–17].

Percutaneous TCC of RSOV is the treatment option in patients who are unfit to undergo bypass, with mild or no aortic regurgitation, simple associated defects (muscular ventricular septal defects, secundum atrial septal defects and small patent ductus arteriosus) and suitable for device closure [18]. Potential Complications during TCC are procedure related aortic regurgitation, failure to deploy the device, encroachment of the device on coronary arteries, residual shunt and device embolisation. The long-term follow-up of TCC patients is mandatory to ensure complete closure of the aneurysm.

CONCLUSION

Transcatheter closure of ruptured SVA is a safe and effective alternative to surgical repair in appropriately selected cases and long-term follow-up is mandatory to ensure complete closure of the aneurysm.

CONFLICT OF INTEREST

“The author(s) declare(s) that there is no conflict of interest regarding the publication of this paper.”

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