Percutaneous Device Closure of Ruptured Sinus of Valsalva Aneurysm in a Patient with Down Syndrome - A Case Report

Jegadeesh J Jayaraman
Department of Cardiology,
Madras Medical College and Government General Hospital

Abstract:
We report the trans-catheter closure of ruptured sinus of Valsalva aneurysm (RSVA) in a female patient aged 18 years. The patient is a known case of Down's syndrome evaluated previously. The patient presented with Shortness of breath of 3 months duration had a continuous murmur on auscultation and was found to have rupture of sinus of Valsalva aneurysm. The site of rupture was from noncoronary cusp into right atrium. Trans-catheter device closure of RSVA was planned. The RSVA was entered from the aorta, and an arteriovenous loop was created, and the Co-coon PDA ductal occluder was used for the closure of ruptured sinus of Valsalva Aneurysm. Complete closure of the RSVA was confirmed. No complications were observed during 6 months of follow-up. Percutaneous closure of a ruptured sinus of Valsalva aneurysm is feasible and can be used as an alternative to surgery in optimally selected patients.

Keyword:
percutaneous closure, ruptured sinus of Valsalva aneurysm, ductal occluder

INTRODUCTION:
Ruptured sinus of Valsalva aneurysm is a rare but well-described clinical entity. Sinus of Valsalva aneurysm is five times more common in Asians in whom it presents typically in adolescence and young adulthood. Percutaneous closure of ruptured sinus Valsalva aneurysm (RSVA) was first attempted in 1994 using a Rashkind umbrella. Since then a few reports have been published with the use of different available closure devices. We would like to share our experience in transcatheter closure of ruptured sinus of valsalva aneurysm in a 18 year old female patient a known case of Down’s syndrome.

CASE REPORT:
A 18 year old female a known case of Down’s syndrome was brought to Cardiology department with complaints of shortness of breath for 3 months duration and easy fatiguability. The breathlessness was NYHA class III at the time of presentation.
On examination she had bounding pulses, blood pressure of 110/60 mmHg and a continuous murmur of Grade 3/6 intensity in the left lower sternal border. ECG showed sinus tachycardia and X-ray chest showed cardiomegaly. Echocardiogram showed situs solitus, Left aortic arch and AV and VA concordance. A fistulous tract was found between the Non-coronary sinus and Right atrium with a wind-sock like appearance confirming the diagnosis of Ruptured sinus of Valsalva aneurysm of Non coronary sinus into the Right atrium. Doppler examination revealed continuous flow across the aneurysm from the aorta into the RA. There was no associated VSD or other congenital defects. Patient had mild pulmonary arterial hypertension and Normal Left Ventricular function .Infective Endocarditis was ruled out. The maximum size of the defect was 8 mm in the aortic end. Trans-esophageal echocardiogram was done to confirm the findings. The treatment options for ruptured sinus of Valsalva aneurysm were discussed with the patient’s parents and an informed consent were obtained. Trans-catheter device closure of Ruptured sinus of Valsalva aneurysm was planned. Under local anesthesia, using Seldinger technique the right femoral artery and right femoral vein were accessed and cannulated using a 6F and 7F sheath respectively. Unfractionated heparin (100 U/kg body weight) was intravenously administered. Routine right and left heart catheterization was performed. A 5 Fr Pigtail catheter was then introduced to perform left ventriculography to screen for Ventricular Septal defect. Aortography was done in Left anterior oblique view, Right anterior oblique view and Anteroposterior views. It was confirmed that no Aortic regurgitation was present. The narrowest diameter of the ruptured sinus of Valsalva aneurysm was measured and it was found to be 8 mm wide and aortography revealed the dye passing freely from the aorta into the Right atrium. A 6 Fr right coronary artery catheter was advanced into the ascending aorta, over a 0.035 angled-tip guidewire (Terumo). This catheter was manipulated to enter into the aneurysm and passed into the right atrium. The guidewire was then advanced into the Inferior vena cava and an Amplatz gooseneck snare was introduced through the femoral venous sheath and the guide wire was snared out and exteriorised through the femoral vein and a stable arteriovenous guidewire loop was thus established. According to the measurement of the aortic end and the point of rupture in the aortogram, a Cocoon duct occluder 2-4 mm larger than the aortic diameter of the ruptured site was chosen to close the defect. The size of the ductal occluder chosen was the 10/12mm PDA ductal occluder. The occluder loaded over its delivery sheath was then introduced from the venous side over the guidewire loop and was positioned across the defect. The entire assembly was pulled back until the larger disk of the device blocked the aortic end of the ruptured site without slipping into the aneurysm. The positioning of the device was confirmed by aortography. After confirming that there was no Aortic regurgitation or Coronary ostial encroachment the device was released from the delivery cable. Finally a check aortogram was done to see the presence of any residual shunt. After the procedure, prophylactic antibiotics were administered for 3 days. The patient underwent TTE before discharge, and received acetylsalicylic acid in the dose of 3-5 mg/kg on a daily basis for 6 months. The patient was followed up for 6 months. There were no complications like infective endocarditis, thromboembolism ,hemolysis or device embolisation.
**Discussion:**
Ruptured sinus of Valsalva aneurysm is a rare cardiac anomaly constituting less than 1% of congenital anomalies of heart and circulation. or right atrium. Although Valsalva aneurysms may involve all 3 aortic sinuses, more frequently the Right and Non-coronary sinus are involved and 2/3rd of the aneurysms arise in the right coronary sinus and 1/4th of aneurysms arise from the non-coronary sinus. The left sinus is not derived embryologically from bulbar septum and therefore is rarely affected by congenital lesions. During rupture they open most frequently to the Right ventricle. Rare sites of rupture include pulmonary artery, LV, LA and pericardial cavity. Sinus of Valsalva aneurysms are usually single, predominating in males about 75%, 30-50% are associated with VSD (especially outlet VSD), proportion of VSD higher if the Right sinus is involved. Prevalence is more in Asian countries. The incidence of associated VSD is also higher in eastern countries. The supracristal VSD (type I) is the predominant type seen in Asian countries, whereas perimembranous VSD (type II) is often seen in western countries.

Patients experience symptoms when the aneurysm ruptures into a cardiac chamber causing continuous murmur, exercise intolerance, symptomatic heart failure, or sudden death, depending on the quantum of the left to right shunt. When it ruptures, mostly to the right heart resulting in left-to-right shunt, the patients may experience severe heart failure.

The usual treatment for ruptured sinus of Valsalva aneurysms is surgical treatment consisting excision of the aneurysmal sac and patch closure of the defect under cardiopulmonary bypass. Although conventional surgical correction under cardiopulmonary bypass carries low mortality, postoperative septicemia, infective endocarditis, and prolonged recovery time make percutaneous device closure an attractive alternative. Cullen et al attempted the first percutaneous closure of RSVA using Rashkind umbrella in 1994.

Since then, Gianturco coil, Amplatzer PDA occluders (ADO) 3, 4, 7, and Amplatzer VSD occluders have been applied in clinical practice. Because of the potential arterial damage using that route, Rashkind umbrella and Gianturco coil failed to be widely used. ADO by venous route have been recommended for ruptured SVA with diameters < 12mm. An arteriovenous loop was applied because it gave the advantage to deliver the transport sheaths through the vein to avoid injuries to the arteries. The coronary ostia are usually quite high in relation to sinus of Valsalva aneurysm. Device impingement occurs rarely. We did not encounter any complications.

**CONCLUSION:**
Percutaneous trans-catheter closure is an attractive option for the treatment of ruptured sinus of Valsalva aneurysms. Since conventional surgery needs cardiopulmonary bypass, the patient is at risk for the development of neurological sequelae, percutaneous closure helps to avoid this cardiopulmonary bypass procedure thereby reducing hospital stay and morbidity. Percutaneous device closure of ruptured sinus of Valsalva aneurysm, as indicated by the encouraging short- and mid-term outcomes, can be feasible and effective in treating optimally selected patients.
Figure 1: Short axis view at the Aortic valve level.

Figure 2: Colour Doppler at Aortic valve level showing L-R shunt from Aorta to RA. Figure 3: Measurement of diameter of aneurysm at Aortic end. Figure 4: Apical four chamber view showing the RSOV to RA.

Figure 5: Continuous wave Doppler across RSOV showing high velocity continuous flow.
Figure 6: Aortogram in LAO cranial showing the RSOV and its measurement

Figure 7: Arterial side guidewire being passed through the RSOV and snared out through the venous side

Figure 8: The ductal occluder being positioned across the RSOV site

Figure 9: The duct occluder device being deployed and is still attached to the delivery cable.

Figure 12: Post procedure echo shows no leak across device
REFERENCES:


