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CASE REPORT
Cardiac imaging

Multimodality delineation of a fistulous ruptured sinus of Valsalva aneurysm: a teaching case report

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Background Ruptured sinus of Valsalva (SOV) is a rare cardiac anomaly with poor prognosis if untreated. Early diagnosis with accurate delineation of its anatomy is critical for timely treatment and choice of surgical vs. percutaneous intervention. Here we report a case of fistulous rupture of SOV; the preoperative multimodality studies including echocardiography, cardiac magnetic resonance and cardiac catheterization provided teaching and learning points.

Case summary A 48-year-old man with history of heart murmur and hypertension presented with a 5-day history of shortness of breath and peripheral oedema. He was diagnosed with rapid atrial flutter. The transthoracic and transesophageal echocardiography showed severe biventricular systolic dysfunction with a left-to-right shunt from ruptured SOV. The colour Doppler by transthoracic and transesophageal echocardiography and cardiac magnetic resonance revealed a swaying shunt flow exiting in direction to the right atrium (RA) and basal right ventricle (RV) during systole and diastole with no myocardial scaring. The left and right heart catheterization showed elevated right-sided pressures, pulmonary capillary wedge pressure, and left ventricular end-diastolic pressure. There was no difference in O2 saturation between venae cavae and RA but a misleading step-up in O2 saturation between RA and RV. Owing to rupture anatomy with uncertainty, the patient underwent surgical intervention. The ruptured SOV tunnelled through the base of tricuspid annulus to the RA very close to the basal RV.

Discussion Even with multimodality studies it can still be challenging to delineate the anatomy of a ruptured SOV without uncertainty preoperatively.

Keywords Sinus of Valsalva • Echocardiography • Cardiac magnetic resonance • Heart failure • Cardiac catheterization • Case report

ESC Curriculum 2.1 Imaging modalities • 2.2 Echocardiography • 2.3 Cardiac magnetic resonance • 7.5 Cardiac surgery • 6.2 Heart failure with reduced ejection fraction

Learning points

- Anatomic delineation of a ruptured sinus of Valsalva (SOV) is critical in deciding the surgical vs. transcatheter approach in treatment and transthoracic and transesophageal echocardiography are the primary modalities in its diagnosis.
- Multimodality imaging including transthoracic and transesophageal echocardiography, cardiac magnetic resonance, and cardiac catheterization are complimentary, but it can still be challenging to define with certainty the exit chamber or potential coexisting defects of a fistulous ruptured SOV.

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Introduction

Ruptured sinus of Valsalva (SOV) is a rare cardiac anomaly and more often causes left-to-right shunt following the path of least resistance. If uncorrected, rupture of the SOV ends up with heart failure (HF) and poor prognosis. Although there are no recent data on the natural history of untreated SOV rupture due to its rarity and that most patients nowadays undergo surgical or percutaneous intervention shortly after its diagnosis, it is generally believed that untreated patients have a survival period of a few months to a few years.[1,2] Therefore early diagnosis with accurate delineation of ruptured SOV anatomy is critical for its timely treatment and choice of surgical vs. percutaneous intervention.[3,4] Here we report a case of a patient presenting with acute HF and rapid atrial flutter (AFlut), found to have fistulous rupture of congenital SOV aneurysm tunnelled into the right atrium (RA). The preoperative delineation of the anomaly was not without uncertainty even with multimodality studies including transthoracic (TTE) and transesophageal echocardiography (TEE), cardiac magnetic resonance (CMR), and left and right heart catheterization, which we believe provided good teaching and learning points.

Timeline

Time from initial presentation	Initial presentation, investigation, and management
Day 1	A 48-year-old man presented with a 5-day history of shortness of breath and peripheral oedema.
	Electrocardiogram showed newly diagnosed rapid atrial flutter.
	Transthoracic echocardiography showed severely reduced left ventricular ejection fraction (LVEF) of 30% and abnormal intracardiac communication suggestive of ruptured sinus of Valsalva (SOV).
Day 2	Transesophageal echocardiography showed no valvular vegetation and confirmed ruptured SOV from non-coronary sinus with a flow swaying in direction to the right atrium (RA) and right ventricle (RV), with uncertainty in exit chamber and questionable coexisting membranous ventricular septal defect.
	Coronary angiography showed no coronary artery disease.
	Left heart catheterization showed elevated left ventricular end-diastolic pressure.
	Aortic root angiogram showed fistulous ruptured SOV.
	Right heart catheterization showed elevated RA pressure, moderate pulmonary hypertension, and elevated pulmonary capillary wedge pressure.
	Oximetry study showed a step-up in oxygen saturations from the RA to RV but not from venae cavae to RA.
Day 4	Cardiac magnetic resonance showed a swaying regurgitant flow from ruptured non-coronary SOV to RA and basal RV in association with cardiac cycle, biventricular systolic dysfunction (LVEF of 23%), and no myocardial late gadolinium enhancement.
Day 7	Atrial flutter converted to sinus rhythm with direct current cardioversion.
Day 26	Surgical repair of fistulous rupture of non-coronary SOV into the RA coursing through the base of the septal leaflet of the tricuspid valve.
Day 71(1.5 months after surgical repair)	Repeat transesophageal echocardiography showed improved LVEF to 40–45%.

Case presentation

A 48-year-old man with a medical history of heart murmur for years and hypertension presented with a 5-day history of shortness of breath and peripheral oedema. He denied chest pain. He was afebrile. The blood pressure was 140/63 mmHg, heart rate 127 beats per minute, respiratory rate 20 breaths per minute, and oxygen (O2) saturation 98% on room air. Cardiopulmonary examination showed irregular heart rhythm with a 2/6 systolic murmur over the right upper sternal boarder and no audible diastolic murmur. Lungs were clear to auscultation. He had 1 + bilateral leg pitting oedema.

The presenting electrocardiogram (ECG) showed AFlut with right axis deviation and rapid ventricular response (*Figure 1*). The complete blood count, metabolic panel, and serial cardiac troponin I levels were unremarkable. The *N*-terminal pro-B-type natriuretic peptide level was elevated at 2405 pg/mL (reference range: 0.0–300.0 pg/mL). The chest X-ray showed cardiomegaly, bibasilar opacity, and small bilateral pleural effusions. The TTE showed severe left ventricular global hypokinesis with left ventricular ejection fraction (LVEF) estimated 30% (see Supplementary material online, *Video S1*). There was trace mitral regurgitation and trivial aortic regurgitation but no significant valve abnormality otherwise. Heart failure with reduced ejection fraction (HFrEF) was diagnosed. Of note, a left-to-right shunting was noted by colour Doppler imaging, pointing to a ruptured SOV and possibly a coexisting perimembranous ventricular septal defect (VSD) (*Figures 2A* and *2B*, Supplementary material online, In-line *video S1*). The left-to-right jet was biphasic during both systole and diastole, with maximal flow velocity of 4.5 m/s on TTE Doppler study (*Figures 2C* and *2D*). The TEE showed normal aortic valve with no vegetation but a left-to-right jet originating from the non-coronary sinus (*Figures 2E* and *2F*, Supplementary material online, In-line *video S2*). The coloured shunt flow on TTE and TEE was dynamic with change in its direction during cardiac cycle suggestive of an exit of the shunt to the RA and possibly to the basal right ventricle (RV) as well.

With a view of interventional management, we performed left heart catheterization and right heart catheterization (RHC), coronary angiography, as well as CMR to evaluate the hemodynamics, rule out significant coronary artery disease, delineate the shunt course, and investigate potential coexisting congenital defect or other myocardial diseases. There was no angiographically significant coronary artery disease (CAD) (*Figure 3*). The left ventricular end-diastolic pressure was elevated (18 mmHg). Aortic root angiography showed a biphasic inverse Y-shaped regurgitant jet from the SOV to the RA and/or the

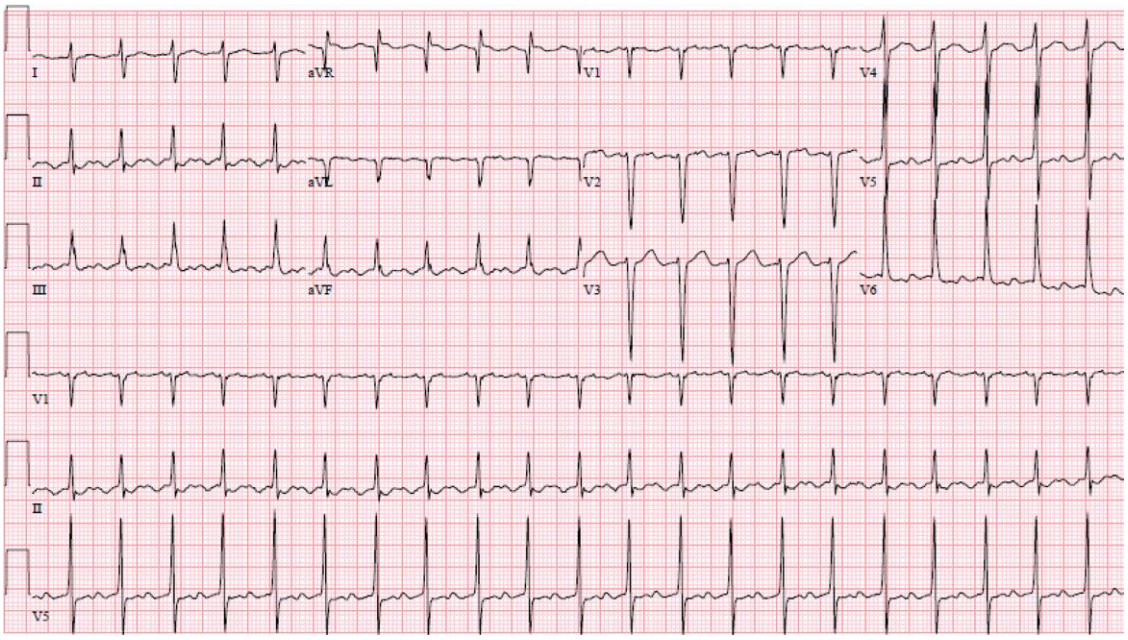


Figure 1 Electrocardiogram at presentation.

RV (see Supplementary material online, *Video S2*). The RHC showed elevated pulmonary arterial pressure of 43/22 mmHg (mean 32 mmHg). The RA pressure was elevated at 14 mmHg and the pulmonary artery (PA) wedge pressure was also elevated at 18 mmHg. Oximetry study for shunt evaluation showed O2 saturations of 74, 74.8, 74, 79, and 79%, in the inferior vena cava (IVC), superior vena cava (SVC), RA, RV, and the PA, respectively, which revealed a step-up in O2 saturation from RA to RV suggesting the presence of VSD. CMR showed severe biventricular systolic dysfunction with LVEF calculated 23% (see Supplementary material online, *Video S3*). On CMR cine sequences, there was a high velocity jet from the non-coronary sinus, which in most views appeared to be coursing along the ventricular side of the septal leaflet of the tricuspid valve and, in some views, along the atrial side of the septal leaflet of the tricuspid valve to the RA and RV (*Figure 4A* and *4B*, Supplementary material online, In-line *video S3*). The ratio of total pulmonary blood flow to total systemic blood flow, Qp:Qs, by CMR volumetric analysis was 2.3. There was no significant myocardial late gadolinium enhancement (LGE) (*Figure 4C*).

The patient underwent electrical cardioversion for the AFlut and apixaban was used for thromboembolism prophylaxis. He underwent open heart surgery and a ruptured congenital SOV aneurysm was diagnosed. The aortic valve was normal in appearance. There was an ~6 mm oval-shaped defect at the base of non-coronary cusp (*Figure 5A*). Through right atriotomy and aortotomy, an aorto-cameral fistula from non-coronary sinus of Valsalva (SOV) to RA was identified coursing through the base of the septal leaflet near its junction with the anterior leaflet of the tricuspid valve, as projected on the root aortogram (*Figure 5B*). Autologous pericardial patch was used to repair the ruptured SOV from both aortic and atrial sides.

LVEF improved to 40–45% by TTE 1.5 months after surgery. The patient has not had recurrent HF since. Of note, during later follow-up, the patient was found to have recurrent paroxysmal AFlut, for which he underwent radiofrequency catheter ablation with a cavotricuspid isthmus line 3 months after the surgery and he has remained in sinus rhythm since. He has been followed up regularly by our cardiology service since the surgical repair of the ruptured SOV. He has been clinically stable and in sinus rhythm in the last more than 3 years.

Discussion

We here presented a teaching case of a patient with fistulous rupture of SOV aneurysm to the RA. Even multimodality preoperative investigation could not conclude in terms of the course or exact exit chamber of the rupture.

Most SOV aneurysms develop from the right coronary sinus followed by non-coronary sinus and much less from the left sinuses,[5,6] and SOV ruptures most commonly into the RV followed by RA.[7–10] VSD is the most common accompanying cardiac defect in patients with SOV aneurysm. Some studies showed that nearly half of the cases with ruptured SOV aneurysm have coexisting VSD.[6–8,10] A perimembranous VSD close to a SOV aneurysm can be misdiagnosed as a ruptured SOV.[11] Embryologically, the right and non-coronary sinuses are believed to derive from the distal bulbar septum, which is also believed to account for a higher prevalence of VSD accompanying the right and non-coronary SOV aneurysm.[2,12] Even with extensive preoperative studies, the VSD can still be missed or misdiagnosed.

The preoperative imaging including TTE, TEE, catheter aortic root angiography, and CMR showed a swaying regurgitant jet from the non-coronary cusp in direction to the RA and RV. Furthermore, the RHC oximetry study showed no significant difference in O2 saturation between IVC/SVC and RA but a step-up in O2 saturation between RA and RV instead. These findings misled us to believe that the SOV aneurysm could have ruptured into the RV or there was a coexistent perimembranous VSD. However, the Doppler signal pattern of the shunt jet indicated a high velocity biphasic left-right flow, which differentiated from an isolated VSD where the left-to-right shunt flow is systolic. Owing to the very close proximity of the rupture exit to the tricuspid annulus the fistulous regurgitant flow swayed into the RA and basal RV during systole and diastole. The shunted blood was directed mostly into the RV leading to a step-up in O2 saturation from RA to RV.

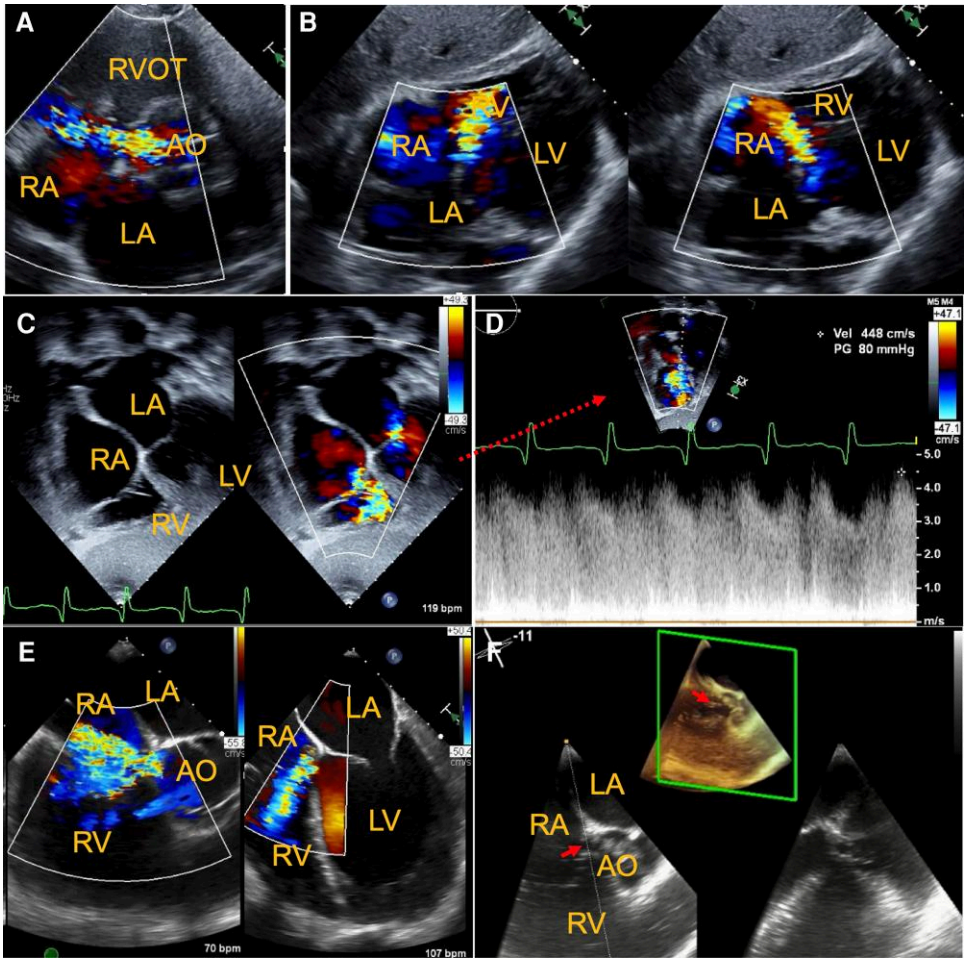


Figure 2 Echocardiographic images. TTE images in parasternal short axis view at aortic valve level (*A*) and subcostal four-chamber view (*B*) showed a left-to-right jet swaying in direction during cardiac cycle. Doppler study using paediatric probe in modified apical four-chamber view showed a biphasic high velocity left-to-right flow (*C, D*).Two-dimensional TEE midesophageal aortic valve short axis (*E*, left) and midesophageal four-chamber views (E, right), as well as three-dimensional TEE view of the aortic valves (*F*) showed non-coronary SOV rupture (red arrows) with a left-to-right flow swaying into RA and RV (*E, F*). RA, right atrium; RV, right ventricle; LA, left atrium; LV, left ventricle; AO, aorta; RVOT, RV outflow tract; SOV, sinus of Valsalva.

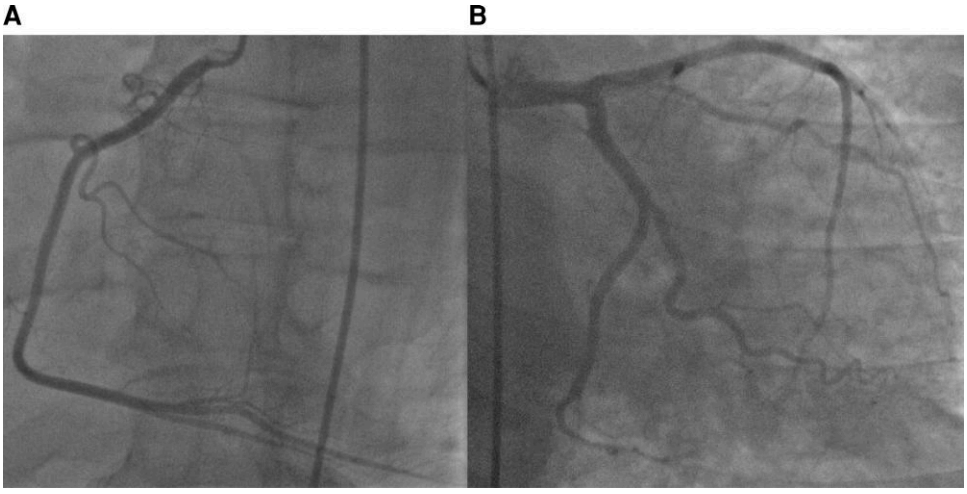


Figure 3 Coronary angiography showing no coronary artery disease. (*A*) right coronary artery; (*B*) left coronary artery.

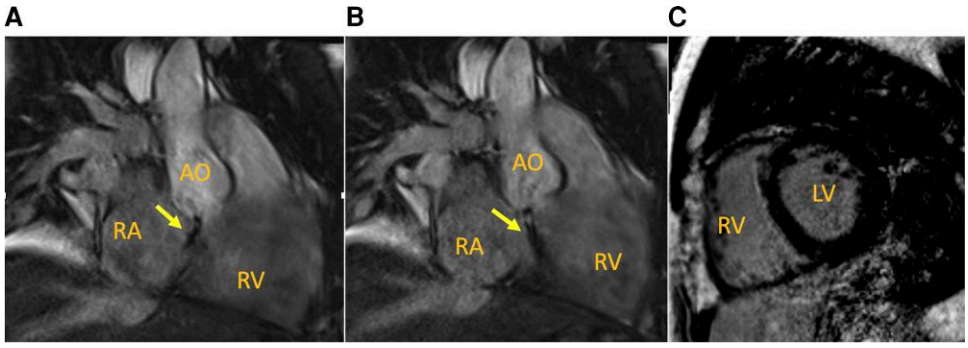


Figure 4 CMR images in RV inflow/outflow view showing a fistulous jet from ruptured SOV to RA during systole and to RV during diastole (yellow arrows) (*A*, *B*) and no significant myocardial late gadolinium enhancement by phase sensitive inversion recovery sequence as seen in a representative midventricular short axis view (*C*). Abbreviations as in legends mentioned in figure 2.

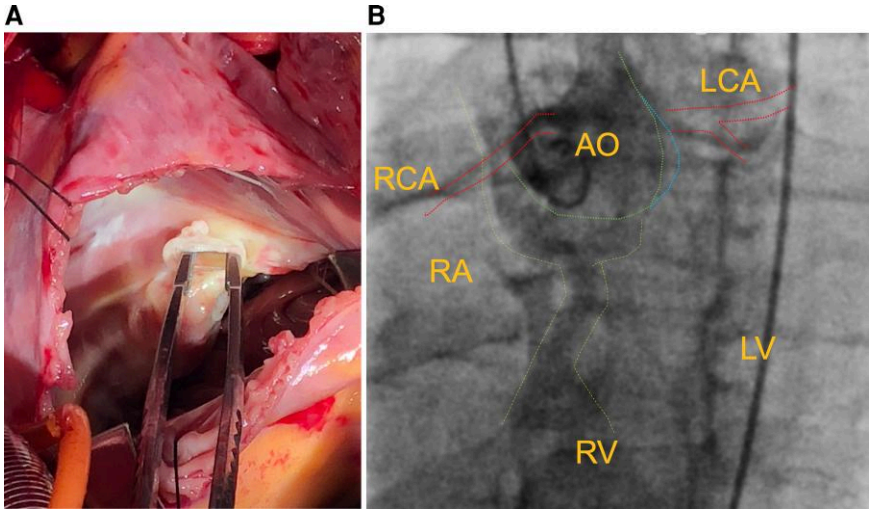


Figure 5 Surgical findings. An atriotomy surgical view of the fistulous ruptured SOV exit to the RA (opened by forceps) (*A*) as projected on the aortography (*B*) shows a linear jet from aortic root into the RA/RV in left anterior oblique view. LCA, left coronary artery; RCA, right coronary artery. Other abbreviations as in legends mentioned in figure 2.

Heart murmur, both systolic and diastolic, can be heard in the setting of unruptured SOV aneurysm.[13] In the setting of SOV rupture, the heart murmur is usually continuous, but the murmur can occur in systolic or diastolic phase.[14] In our case, the patient has a known long-standing history of heart murmur which could be from the enlarging SOV aneurysm related local blood flow alterations or turbulence. At this presentation with ruptured SOV, the physical exam revealed a systolic murmur over the upper sternal boarder, but we did not appreciate diastolic murmur. This is consistent with the literatures. The lack of appreciable diastolic murmur could also be due to the relative less pressure gradient across the rupture during the diastolic phase than systolic phase, or due to similar factors affecting the audibility of diastolic murmur in the setting of aortic regurgitation.[15]

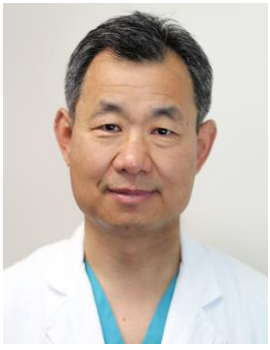
The patient presented with biventricular failure as demonstrated by echocardiography, CMR, and cardiac catheterization. The coronary angiography showed no CAD and CMR showed no LGE to indicate myocardial fibrosis. Therefore, the biventricular failure could be attributed to the hemodynamic deterioration from ruptured SOV, which also contributed to the development of AFlut with tachycardia. Tachycardia in turn further worsened cardiomyopathy and HFrEF.

So far there is no guideline regarding the treatment of ruptured SOV in terms of surgical vs. transcatheter approach. Surgical repair remains the treatment of choice for ruptured SOV especially in cases with significant aortic regurgitation and more complex associated lesions such as endocarditis, bicuspid aortic valve, tunnel-type fistulous connections, larger defect size, and multiple site of rupture, but transcatheter device closure has emerged as an alternative option for patients with suitable anatomy especially with advent of new devices.[16]

In summary, we presented a teaching case of multimodality cardiac evaluation for a ruptured congenital SOV aneurysm from non-coronary sinus with a fistulous course exiting into the RA. The resultant shunt jet swayed in direction to the RA and RV during cardiac cycle, due to its very close proximity to the basal RV. Preoperative anatomic delineation of a ruptured SOV is critical in deciding surgical vs. transcatheter

intervention, and it can be challenging even with multimodality studies. In our case, transcatheter approach was not considered for treatment due to the fistulous course of the ruptured SOV with uncertainty in terms of the exit chamber of the shunt and potential coexisting defect VSD.

Lead author biography



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Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in accordance with COPE guidelines.

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