



## Case Report

**Doubly Committed Subarterial Ventricular Septal Defect Coexisted with Ruptured Aneurysm of the Right Sinus Valsava**Irma Maghfirah<sup>1\*</sup>, Rochmad Romdoni<sup>1,2</sup><sup>1</sup>Department of Cardiology and Vascular Medicine, RSUD Dr. Soetomo, Surabaya, Indonesia.<sup>2</sup>Faculty of Medicine, Universitas Airlangga, Indonesia.

## ARTICLE INFO

*Article history:*

Submitted December 2020

Reviewed January-June 2021

Revised January-February 2021

Accepted March 2022

Available online March 2022

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*Keywords:*

Aneurysm of sinus valsava

Ventricular septal defect

## ABSTRACT

Sinus of Valsalva aneurysms (SVAs) are uncommon cardiac abnormalities and most of them are in congenital origin. Some patients may have SVAs that concomitant with ventricular septal defect (VSD). Here, we reported case of a 37-year-old male presented with worsening exertional dyspnoea. Thrills and loud continuous murmur along the left sternal border were discovered during physical examination. Echocardiogram displayed ruptured sinus of valsalva aneurysm to the right ventricle along with supracristal ventricular septal defect (VSD). The patient underwent surgical correction with patch repair and aortic valve replacement. Most cases of SVAs were originated from right coronary sinus. They usually ruptured into right ventricle. Ventricular septal defects often coexisted with this condition. Echocardiography provided a complete evaluation of such cases and prohibited missed diagnosis of other coexistent congenital heart defects.

**Introduction**

Sinus of Valsalva aneurysms (SVAs) are very rare cardiac abnormalities. They can be classified into two groups based on their etiology, congenital and acquired. The congenital cases are more common than the acquired ones. They are caused by the weakening of structure located between annulus fibrosus and aortic media<sup>[1]</sup>. More than half cases of SVAs originated in the right coronary sinus, followed by non-coronary sinus and rarely left coronary sinus<sup>[2]</sup>. The patient with SVA usually remains asymptomatic unless it is complicated by a ruptured. Ruptured aneurysm of Sinus of Valsalva is highly associated with other cardiac lesion, especially ventricular septal defects, bicuspid aortic

valve, and aortic regurgitation<sup>[3]</sup>. Echocardiography remains the mainstay diagnostic tool for such cases. It helped the physician to establish the diagnosis of SVA ruptured as well as VSD<sup>[4]</sup>. We report a patient with SVA that originated from right coronary sinus which is later ruptured into the right ventricle along with supracristal ventricular septal defect that underwent surgical successful operation.

**Case Presentation**

A 37-year-old man presented to our outpatient clinic with worsening exertional dyspnea over previous 6 months. Shortness of breath is complained first

when he did vigorous activity, but since 2 months, its intensity increased, as the symptoms often appeared when patient underwent moderate to low intensity activities. Physical examination revealed loud superficial continuous murmur and thrills along left sternal border. Other signs and symptom of right heart failure like ascites and slight pitting oedema of inferior extremity were also present. Electrocardiogram revealed sinus tachycardia at a rate of 115 beats/min and T-wave inversion in lateral and inferior leads. Cardiomegaly was shown from chest x-ray along with bilateral cephalization as a sign of mild pulmonary congestion.

Transthoracic echocardiography revealed continuous flow in systole and diastole from aorta to the right ventricle through a ruptured of right sinus valsalva aneurysm. Parasternal short axis view on aortic valve level demonstrated abnormal flow from sinus valsalva to the right ventricle at the 12 to 2 o'clock direction (figure 1). Meanwhile, color Doppler that was obtained from parasternal right ventricular outflow tract view, exhibited ventricular septal defect jet across the supracristal VSD (figure 2). The shunt direction was from left to right. There was another defect existed in interatrial septum, a Patent Foramen Ovale (PFO). Transesophageal echocardiography confirmed the diagnosis of ruptured of sinus valsalva aneurysm (figure 3) along with supracristal VSD (figure 4). The other echocardiographic findings were mild aortic regurgitation, preserved systolic ejection fraction, all chamber dilatation along with mild pulmonary hypertension.

Left heart catheterization detected shunting from sinus valsalva toward right ventricle, consistent with ruptured aneurysm of right coronary sinus of valsalva. On the contrary, ventriculography revealed no defects at interventricular septum, confronting echocardiogram findings.

Patient underwent surgery with cardiopulmonary bypass support in a hypothermic state. The surgical approach used sternotomy method. Cardioplegic was given by hand-held technique directly toward both ostiums of left main and right coronary artery respectively. During the process, supracristal ventricular septal defect was confirmed (figure 5) along with ruptured aneurysm of right coronary sinus of valsalva (figure 6). There were episodes of torsade de pointes and atrial fibrillation. The ventricular septal defect was repaired using a 1.4 cm Gore-Tex patch. Meanwhile, the ruptured sinus valsalva was obliterated using pericardial patch. Patent foramen ovale was left untreated since the patient had pulmonary hypertension. As for the aortic valve, St. Jude prosthesis valve was used to replace it.

Post operative transesophageal echocardiography showed proper site and function of prosthetic aortic valve, with no valvular and paravalvular leakage, along with good function of both pericardial and Gore-tex patches. Patient went through good recovery, dyspnea was diminished, and was discharged 7 days after surgery.

## Discussion

Sinus valsalva aneurysms are rare cardiac lesions. It may be divided based on etiology as two groups, acquired and congenital<sup>[1]</sup>. Men were more affected than women with 3:1 ratio and Asian race bear the highest incident<sup>[3]</sup>. In vast majority of patients, aneurysm of sinus valsalva originated from right coronary sinus, followed by noncoronary sinus. Meanwhile SVA from left coronary sinus are the least common. SVA patients usually remain asymptomatic. Ruptured of right SVA generally occurred at second or third decade of life and most of them ruptured toward right ventricle<sup>[4]</sup>. The signs and symptoms are similar to congestive heart failure. Ruptured SVA frequently coexisted with

other types of cardiac anomalies as well, such as ventricular septal defects and aortic valve disease. Type of VSD highly associated with ruptured of SVA is the supracristal type (type 1 VSD). It is the predominant type in Asian population [5]. In our case, ruptured aneurysm of right sinus valsalva coexisted with supracristal VSD which was detected by echocardiography and confirmed by cardiothoracic surgeon.

Echocardiography examinations, both transthoracic and transesophageal echocardiography, remain the mainstay diagnostic tools to establish the diagnosis of cardiac anomalies, including SVA and VSD. As for SVA ruptured, the accuracy of transthoracic echocardiography is about 75% [6]. It helped us determining not only the the direction of the rupture, but also the size and site of aneurysm. Subsequently, it will guide the physician to discover the coexistent cardiac defects. In this presented case, the patient suffered from right sinus of valsalva aneurysm that ruptured toward right ventricle. Trans thoracic echocardiography displayed discontinuity of right coronary sinus with shunting from aorta to the right ventricle as observed in parasternal short axis views. Cardiac catheterization possessed good sensitivity and sensitivity with regard to SVA rupture as well [5].

In this patient, both TTE and TEE confirmed the presence of supracristal VSD. Still, the catheterization showed otherwise. In case of supracristal ventricular septal defects, compared to catheterization, echocardiography possesses better sensitivity with regard to supracristal VSD. Still, both are highly specific. The sensitivity and specificity of echocardiography in diagnosing supracristal VSD was 99% and 95% respectively [6]. Supracristal VSD, which is also known as doubly committed subarterial VSD, was the most common cardiac lesion accompanying SVA, especially SVA originated from right coronary sinus that ruptured

into right ventricle [5]. It is fundamental for cardiologist to not miss the presence of this type of VSD in SVA patient. Some additional views of echocardiographic examination must be obtained in determining whether supracristal VSD exists or not, especially in patient with ruptured SVA. Parasternal right ventricular outflow tract view, subcostal long axis view, along with subcostal short axis views may help in visualizing jet of VSD [6].

## Conclusion

Ruptured aneurysms of right sinus of Valsalva are rare cardiac anomalies. In most cases it usually ruptured toward right ventricle. They often coexisted with supracristal VSD. Echocardiography provides complete evaluation of these cases and help the physician not to miss in diagnosing these cardiac defects.

## Acknowledgement

There is no conflict of interest.

## References

1. Feldman D, Gade C and Roman M. 2005. Ruptured Aneurysm of the Right Sinus of Valsalva Texas Hear. Inst. J. 32:555–9.
2. Jin Y, Han X, Wang H, Wang Z, Fang M, Yu Y and Fang X. 2017. Coexisting ventricular septal defect affects the features of ruptured sinus of Valsalva aneurysms Saudi Med J. 38:257–61.
3. Moustafa S, Mookadam F, Cooper L and Carolina N. 2007. Sinus of Valsalva Aneurysms — 47 Years of a Single Center Experience and Systematic Overview of Published Reports Am. J. Cardiol. 99:1159–64.
4. Yan F, Huo Q, Qiao J and Murat V. 2008. Surgery for Sinus of Valsalva Aneurysm : 27-Year Experience with 100 Patients Asian Cardiovasc. Thorac. Ann. 16:361–5.

5. Ishii, Mie; Masuoka H, Emi, Yoshiharu; Takuya M, Ito, Masaaki; Nakano T. 2003. Ruptured Aneurysm of the Sinus of Valsalva Coexisting With a Ventricular Septal Defect and Single Coronary Artery Circulation 67:470–2.
6. Dev, Vishva Goswami Kewal, Shrivastava Savitri, Bahl Vinay S A 1993 Echocardiographic diagnosis of aneurysm of the sinus of Valsalva Am. Heart J. 126:930–6.
7. Ott DA. Aneurysm of the Sinus of Valsalva. Seminars in Thoracic and Cardiovascular Ott DA. 2006. Aneurysm of the sinus of Valsalva. Semin Thorac Cardiovasc Pediatr Card Surg Annu Surg.165–176.
8. Moustafa S, Mookadam F, Cooper L, et al. 2007. Sinus of valsalva aneurysms - 47 years of a single center experience and systematic overview of published reports. Am J Cardiol. 99: 1159–1164.
9. Dmitry NF, Gade CL, Roman MJ. 2005. Ruptured Aneurysm of the Right Sinus Valsalva. Tex Heart Inst J 32: 555-9.
10. Yan F, Abudureheman M, Huo Q, Shabiti A, Zhu T, Liu Z. 2014. Surgery for sinus aneurysm from Valsalva: 33-year of a single center experience. Chin Med J (Engl) 127: 4066-4070.
11. Bricker AO, Avutu B, Mohammed TL, et al. 2010. Valsalva sinus aneurysms: findings at CT and MR imaging. Radiographics. 30: 99 - 110.
12. JM Troupis, Nasis A, Pasricha S, et al. 2013. Sinus valsalva aneurysm on cardiac CT angiography: assessment and detection. J Med Imaging Radiat Oncol. 57: 444–447.
13. Weinreich M, Yu PJ, Trost Biana. 2015. Sinus of Valsalva Aneurysms: Review of the Literature and Update on Management. Clin. Cardiol. 38, 3, 185-189.

## Supplementary Data

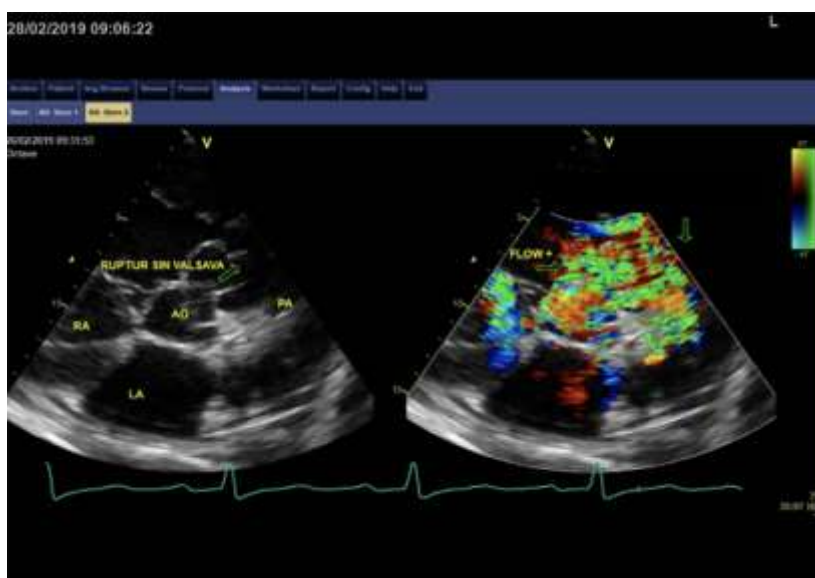


Figure 1. Color Doppler of Trans thoracic echocardiography (parasternal short-axis view). Jet lesion of ruptured right SVA toward right ventricle is displayed (arrow). RA = right atrium; PA = pulmonary artery; LA = left atrium; Ao = aorta

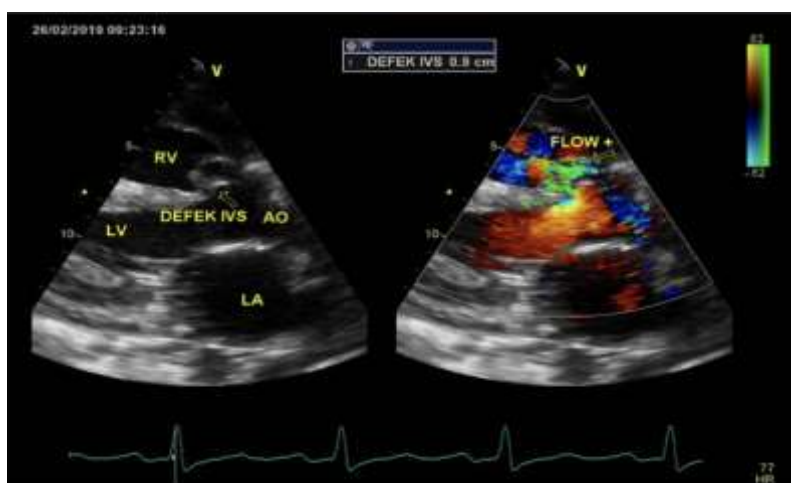


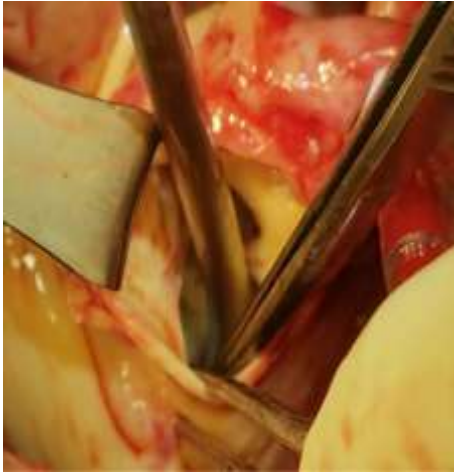
Figure 2. Transthoracic echocardiography (parasternal right ventricular outflow tract view) exhibited ventricular septal defect jet across the suprasternal VSD.



**Figure 3.**  
Transesophageal echocardiography displayed ruptured of SVA toward right ventricle



**Figure 4.**  
Transesophageal echocardiography showed defect in interventricular septum



**Figure 5.** Supracristal ventricular septal defect was confirmed during surgical process (arrow)



**Figure 5.** Ruptured aneurysm of right sinus of Valsalva was displayed (arrow)