# An Unusual Cause of Acute Abdominal Pain and Unexplained Dyspnea in a Young Man: A Sinus of Valsalva Aneurysm

Rahul V. Annabathula, MD, Li Zhou, MD, PhD, Edward H. Kincaid, MD, Richard B. Stacey, MD, Sujethra Vasu, MD, and Bharathi Upadhya, MD, *Winston-Salem, North Carolina*

## INTRODUCTION

A sinus of Valsalva aneurysm (SVA) is defined as an abnormal dilatation of one or more of the aortic sinuses at the site of the aortic valve annulus and the sinotubular junction. Sinus of Valsalva aneurysms are rare, and prevalence ranged from 0.14% to 0.23% in a large surgical series.[1] It is the result of underlying weakness of the elastic lamina between the aortic media and the annulus fibrosis.[2] Unruptured aneurysms can largely be asymptomatic, but a ruptured aneurysm is a deadly complication that can result in a myriad of sequelae depending upon the underlying anatomy of the shunt itself. Here, we present a case of a 25-year-old man who presented with nonspecific symptoms of acute-onset abdominal pain and dyspnea and was diagnosed with a ruptured SVA into the right atrium (RA), leading to an aorto-RA fistula. This case report highlights the importance of a thorough cardiac workup in young patients with nonspecific symptoms, unexplained dyspnea, and a diastolic murmur without any significant comorbidities.

## CASE PRESENTATION

A 25-year-old man with no relevant medical history presented to the emergency department with acute-onset right upper quadrant abdominal pain, nausea, and vomiting over the last 24 hours. The patient had been in good health prior to this presentation. Upon further questioning, the patient also endorsed experiencing dyspnea on exertion over the last few days. Of note, he denied chest pain, orthopnea, fever, or chills. The patient did endorse occasional marijuana use but denied any other illicit substance use.

Upon initial presentation to the emergency department, the patient's vital signs were as follows: temperature 98.7°F, heart rate 106 beats/minute, respiration 16 breaths/minute, blood pressure 122/57 mm Hg, and oxygen saturation 97% on room air. On physical

From the Department of Internal Medicine (R.V.A.); Department of Internal Medicine, Section of Cardiovascular Medicine (L.Z., R.B.S., S.V., B.U.); and Department of Cardiothoracic Surgery (E.H.K.), Wake Forest School of Medicine, Winston-Salem, North Carolina.

Keywords: Sinus of Valsalva aneurysm, Rupture, Echocardiography

Conflicts of Interest: The authors reported no actual or potential conflicts of interest relative to this document.

Correspondence: Rahul V. Annabathula, MD, Department of Internal Medicine, Wake Forest School of Medicine, Medical Center Boulevard, Winston-Salem, North Carolina 27157-1045. (E-mail: *rannabat@wakehealth.edu*).



2468-6441
https://doi.org/10.1016/j.case.2022.02.005

## VIDEO HIGHLIGHTS

**Video 1:** Two-dimensional TTE. Zoomed right ventricular inflow view demonstrates multiple linear, highly mobile, echogenic masses near the tricuspid valve in the RA.

**Video 2:** Two-dimensional TTE PSSAX basal view at the level of the normal appearing aortic and pulmonic valve leaflets demonstrates multiple, highly mobile echogenic masses in the RA at the junction of the septal tricuspid valve leaflet and the right sinus of Valsalva. At this time, the diagnosis of infective endocarditis with TR was made, but a ruptured congenital SVA or VSD could not be entirely excluded, and TEE was recommended.

**Video 3:** Two-dimensional TTE. Zoomed right ventricular inflow color Doppler view demonstrates suspected moderate TR with increased flow across the tricuspid valve during diastole and an atypical mosaic flow pattern emphasizing the heterogeneity of flow.

**Video 4:** Two-dimensional TTE PSSAX color Doppler view demonstrates severe turbulence suggesting TR, a small VSD, or a possible SVA rupture.

**Video 5:** Two-dimensional TEE midesophageal view (0°) rightward rotated right ventricle–focused apical long-axis view without (*left*) and with (*right*) color Doppler. Multiple linear mobile masses are seen in the RA with what appears to be at least moderate TR, raising the concern for tricuspid valve endocarditis.

**Video 6:** Two-dimensional TEE midesophageal view (63°) without (*left*) and with (*right*) color flow Doppler more clearly demonstrates the color flow disturbance to originate with a previously unconfirmed echo-free space in the anterior sinus of Valsalva. Additionally, the source of the mobile tissues seen in the RA with TTE are now seen to conform to a long windsock SVA, which has ruptured and forms a communication between the aorta and the RA. There is no VSD seen nor significant TR on this image.

**Video 7:** Two-dimensional TEE midesophageal view (123°) rightward rotating sweep without (*left*) and with color flow Doppler (*right*), which better demonstrates the origin of the pathologic flow within the right sinus of Valsalva. There is proximal flow acceleration suggesting a large shunt.

**Video 8:** Intraoperative two-dimensional TEE images with color flow Doppler postrepair, midesophageal (64°) view, demonstrate resolution of the SVA.

**View the video content online at www.cvcasejournal.com.**

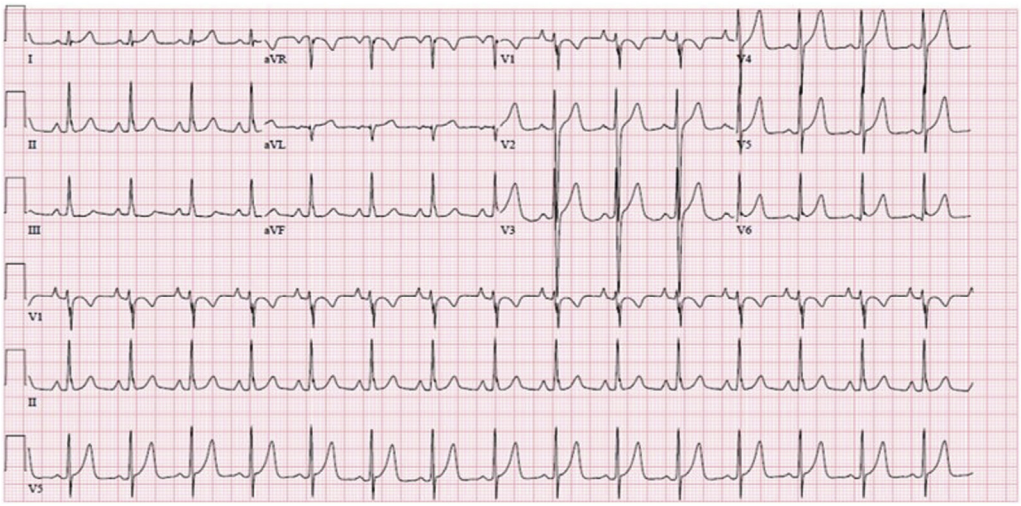


**Figure 1** Twelve-lead electrocardiogram demonstrating sinus tachycardia without pathologic ST changes.

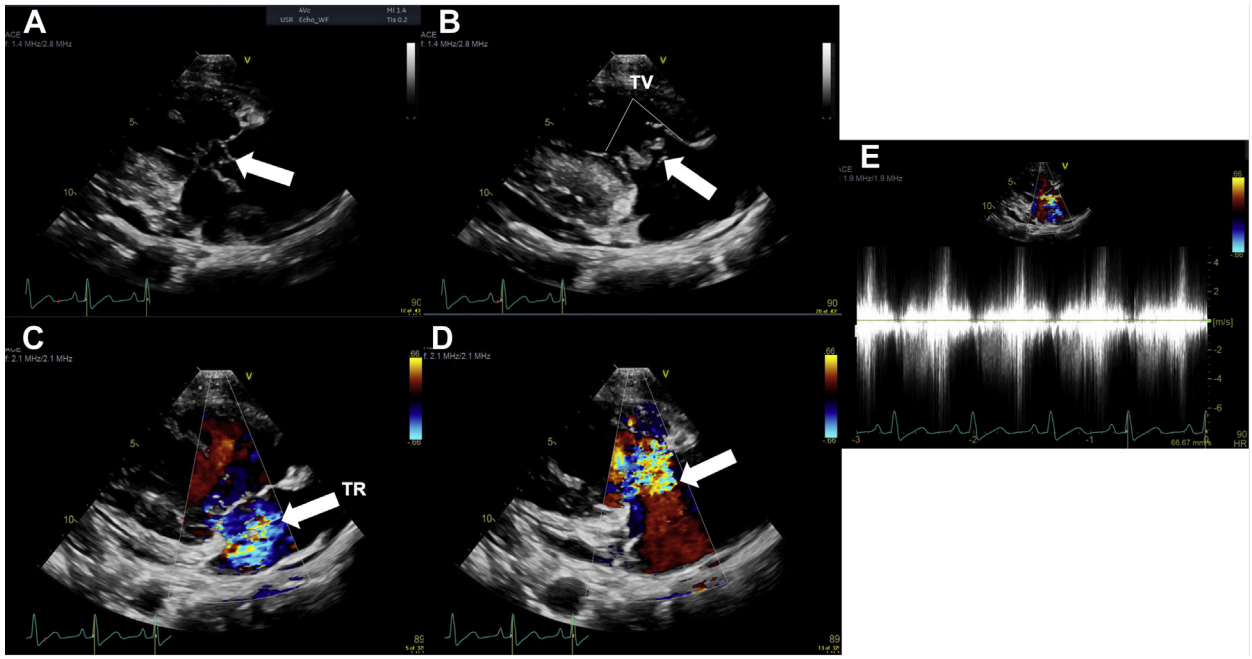


**Figure 2** Two-dimensional TTE zoomed right ventricular inflow view without **(A, B)** and with **(C, D)** color flow Doppler and **(E)** continuous-wave Doppler spectrum. **(A)** Systolic and **(B)** diastolic phases demonstrate multiple linear, mobile, echogenic masses near but possibly not attached to the tricuspid valve in the RA (*arrow*). **(C)** Systolic and **(D)** diastolic phases demonstrate TR and increased turbulent flow across the tricuspid valve despite nonstenotic valve motion (*arrow*). **(E)** A high-velocity flow spectrum is seen throughout the cardiac cycle. The spectral Doppler noise pattern suggests that the cursor is not aligned to flow and the flow origin is out of plane with the image.

examination, the patient was conversant but uncomfortable in general. Cardiac auscultation was notable for normal S1, loud S2 in the pulmonary area, and a holosystolic murmur with diastolic murmur along the left parasternal area. There were no signs of lower extremity edema, and the dorsalis pedis pulse was 2+ bilaterally. Abdominal and lung examinations were unremarkable.

Admission labs were notable for negative SARS-COV-2, Flu A, and Flu B. Initial high-sensitivity troponin was elevated at 31 pg/mL, and B-type natriuretic peptide was elevated at 380 pg/mL. The patient had a leukocytosis at 11.6 $\times$ $10^9$/L and normocytic anemia (hemoglobin, 12.6 g/dL). The urine drug screen was positive for cannabinoids only. Electrolytes were within normal limits. Creatinine was 1.18 mg/dL. Aspartate aminotransferase and alanine aminotransferase were elevated at 168 U/L and 245 U/L, respectively. The hepatitis panel only revealed prior immunization for hepatitis B.

Electrocardiography demonstrated sinus tachycardia without pathologic ST changes (Figure 1). Transthoracic echocardiography (TTE)

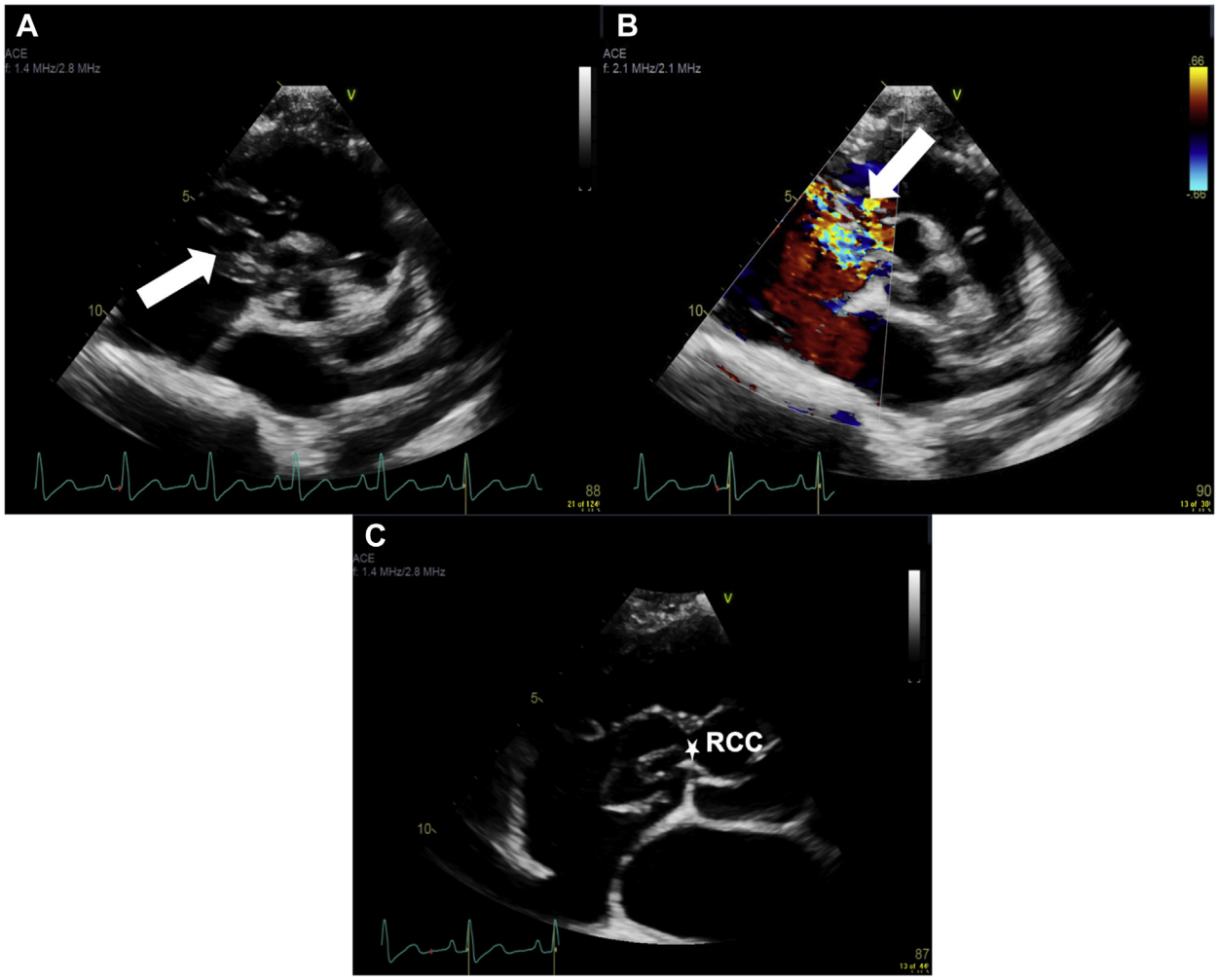


**Figure 3** Two-dimensional TTE Parasternal short-axis basal view without **(A, C)** and with **(B)** color flow Doppler. **(A)** Multiple, highly mobile echogenic masses are seen in the RA at the junction of the septal tricuspid valve leaflet and the right sinus of Valsalva (*arrow*). **(B)** Severe color turbulence within this region is seen during diastole (*arrow*). **(C)** In this zoomed view, suspicion for an SVA (*star*) arising from RCC is demonstrated. *RCC*, Right coronary cusp.

demonstrated multiple linear, highly mobile, echogenic masses near the tricuspid valve in the RA (Figures 2, 3, Videos 1, 2). Color flow Doppler demonstrated an atypical mosaic flow pattern with unclear origin (Figures 2, 3, Videos 3, 4). Continuous-wave Doppler across the tricuspid valve demonstrated high-velocity flow throughout the cardiac cycle (Figure 2). The spectral Doppler noise suggested that the cursor was not aligned to flow. This finding further suggested that the flow origin was out of plane with the image, which was consistent with an eccentric origin and does not suggest either a typical tricuspid regurgitation (TR) or a ventricular septal defect (VSD), although a very eccentric TR jet or serpiginous VSD may look similar. At this time, the diagnosis of infective endocarditis with TR was made, but a ruptured congenital SVA or VSD could not be entirely excluded, and transesophageal echocardiogram (TEE) was recommended.

Transesophageal echocardiogram more clearly demonstrated the origin of the color flow disturbance to originate with a previously unconfirmed echo-free space in the anterior sinus of Valsalva (Figure 4). Additionally, the source of the mobile tissues seen in the RA with TTE was now seen to conform to a long windsock SVA, which had ruptured and formed a communication between the aorta and the RA (Figure 5, Videos 5-7). There is no VSD or significant TR. Three-dimensional TEE images confirmed this (Figure 6). The cardiothoracic surgery team was consulted, and the patient underwent emergent surgery. The ruptured SVA along with the fistulous tract was easily identified during the procedure. The SVA, rupture, and fistula were repaired using a pericardial patch (Figure 5). Repeat intraoperative TTE showed closure of the SVA and resultant fistula (Figure 7, Video 8). The patient's postoperative course was uneventful, and he was discharged on postop day 4.

## DISCUSSION

The majority of SVAs are congenital in nature. Aneurysms of the right and noncoronary sinuses are more common than those of the left sinus. Sinus of Valsalva aneurysms are also often associated with other cardiac abnormalities, such as a bicuspid aortic valve (15%-20%), VSD (30%-60%), and aortic regurgitation (44%-50%).[3] Acquired aneurysms are caused by conditions affecting the aortic wall, such as infections, degenerative diseases (atherosclerosis, connective tissue

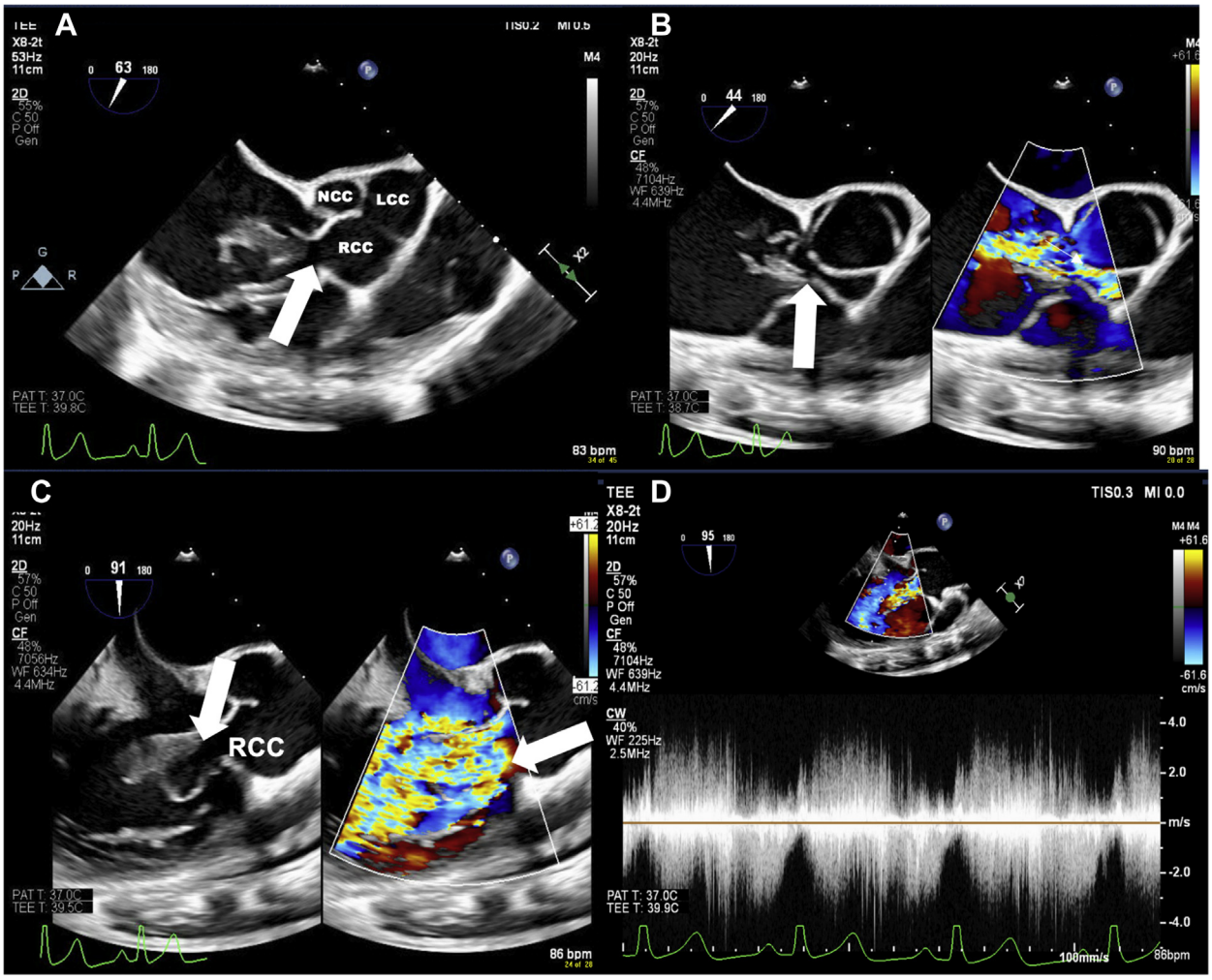


**Figure 4** Two-dimensional TEE midesophageal view (63°) without **(A)** and with **(B, C)** color flow and **(D)** continuous-wave Doppler. **(A)** Each of the aortic valve cusps are well demonstrated in diastolic with the SVA (*arrow*) now well detailed arising from the right coronary cusp (RCC). **(B)** At 44°, the SVA origin and rupture with the aorto-to-right atrial fistula is demonstrated. **(C)** At 91°, the relationship of the SVA (*arrow*) with the aortic root and valve is shown. There is proximal flow acceleration at the origin of the aorto-to-right atrial fistula suggesting a large shunt. **(D)** With the continuous-wave Doppler cursor placed across the tricuspid valve, a noisy high-velocity flow throughout the cardiac cycle is seen. *LCC*, Left coronary cusp; *NCC*, noncoronary cusp; *RCC*, right coronary cusp.

disorders, or cystic medial necrosis), or trauma.[4] Autoimmune diseases have also been associated with the development of SVAs.

Sinus of Valsalva aneurysms often go undetected for many years. Symptoms of SVA rupture include substernal chest pain, abdominal pain, and dyspnea. While our patient's presentation lacked chest pain, the remainder of his presentation remains consistent with conventionally reported symptoms for someone with SVA rupture.[5] Most aneurysms rupture into the right chambers and rarely into the left chambers and pulmonary arteries. Noncoronary sinus aneurysms tend to rupture into the RA, and right coronary sinus aneurysms can rupture into either the RA or right ventricle.[6] Early detection and diagnosis of a ruptured SVA is key to providing prompt intervention and correction. Although not always definitive, the physical exam can afford clinicians some information that can increase suspicion for the underlying diagnosis. In the case of ruptured SVAs, the hallmark sound heard on auscultation is described as a continuous, sawing-like murmur.[5,7] A diastolic decrescendo murmur indicative of aortic regurgitation may also be auscultated.

Given its numerable presentations and proximity to multiple, critical structures, a ruptured SVA poses a diagnostic challenge. There is significant operator variability in the interpretation of both TTE and TEE images. It is the responsibility of the echocardiographer to format images using nonstandard views and different modalities (e.g., two-dimensional, three-dimensional, spectral Doppler, and color flow Doppler) to identify defects that are close to each other. Color Doppler, in the case of ruptured SVAs, affords clinicians the ability to demonstrate continuous flow in both systole and diastole due to the high-pressure system housed within the aorta.[8] As echocardiography is subject to interstudy variability, it raises the importance of case reports to help expand the knowledge base for diagnosing uncommon cardiac pathologies rarely seen outside of high-volume echo laboratories.

Identifying a SVA rupture alone from a SVA rupture with a VSD can be difficult. Furthermore, SVAs that are found near VSDs are often misidentified as ruptured SVAs. An SVA that is associated with aortic regurgitation often obfuscates the diagnosis of an underlying

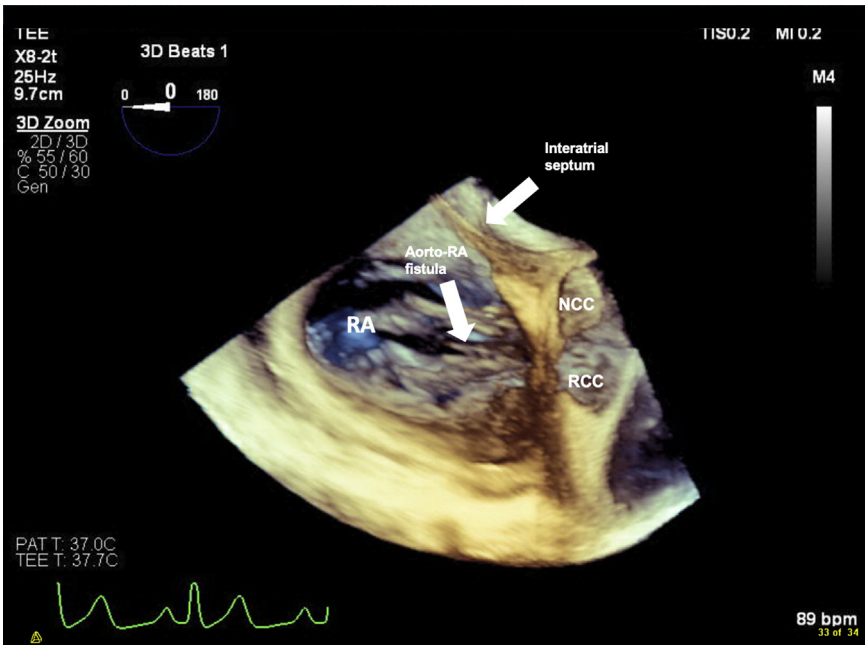


**Figure 5** Three-dimensional volume-rendered TEE showing aorto-RA fistula. *NCC*, Noncoronary cusp; *RCC*, Right coronary cusp.

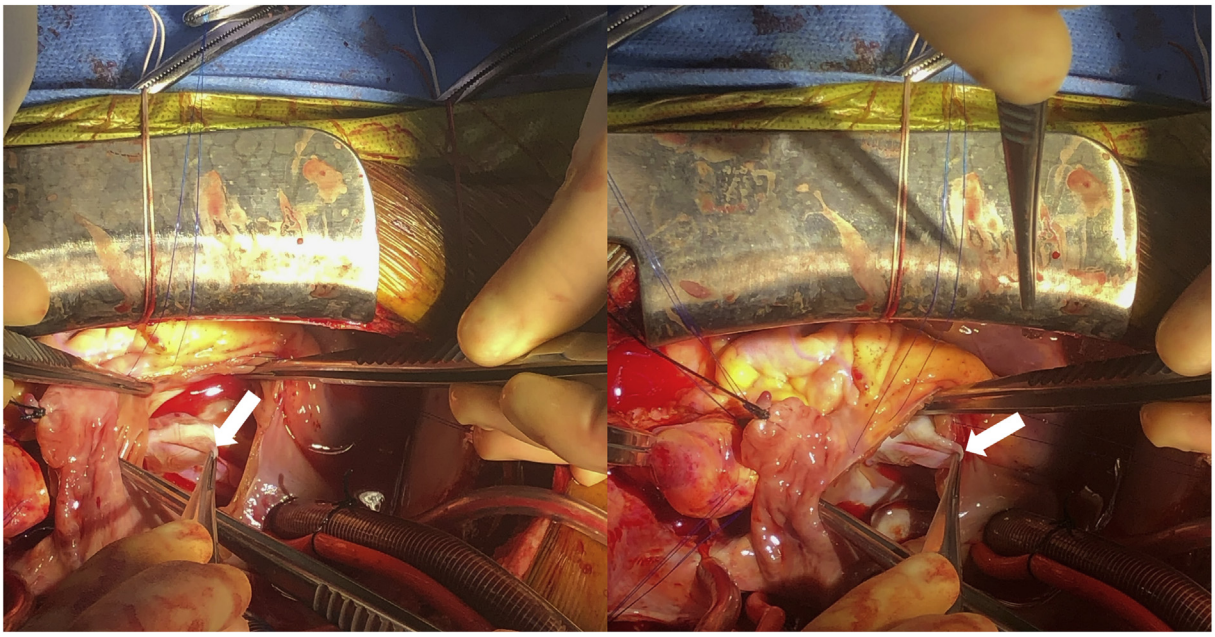

**Figure 6** Surgeon's view of the ruptured SVA. An aortotomy is performed, and the right atrial end of the aneurysmal sac is everted out of the incision. The forceps is pinching the aneurysmal sac protruding into the RA (*arrow*).

VSD.[9] To further delineate a ruptured SVA from a VSD requires precise imaging and the use of spectral Doppler differences of the jets with regard to the annulus of the aortic valve and the exact occurrence within the cardiac cycle.[10]

A single-center study of 212 patients with SVA corroborated TTE findings with surgical findings and found that the sensitivity, specificity, and accuracy of TTE were 93.9%, 99.9%, and 99.8%, respectively.[11] However, since TTE may be poor quality or leave the interpreting physician with doubt regarding the specific underlying pathology, TEE is an appropriate next test to consider, as it affords the ability to acquire higher-quality images and, ultimately, confers the imaging modality with an overall increased accuracy of diagnosis.

Transesophageal echocardiogram is important in surgical planning as left-to-right communications must be promptly identified to allow the modification of the venous drainage technique. If a preoperative TEE is not performed, a comprehensive baseline intraoperative TEE

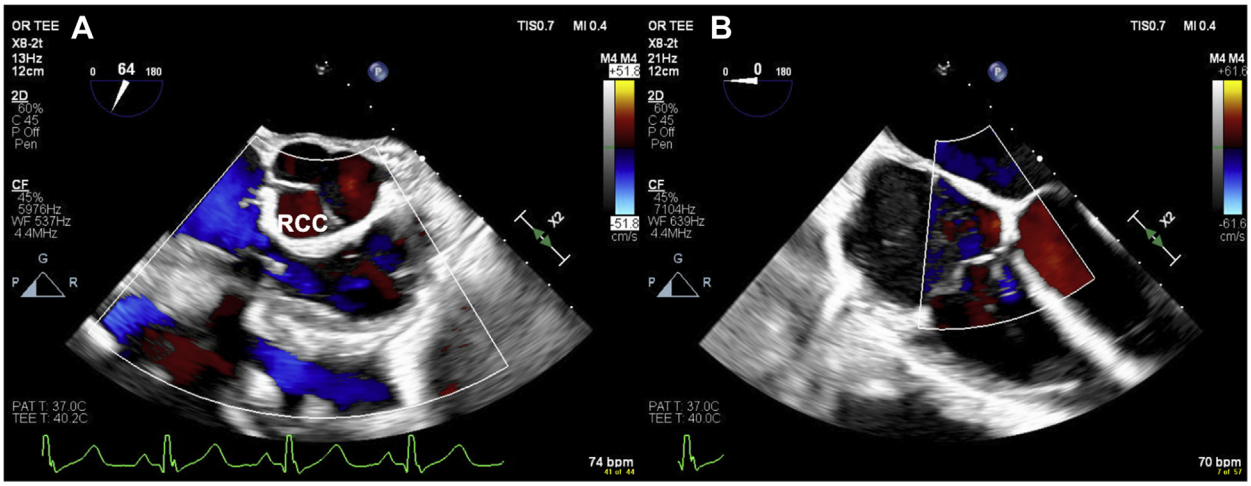


Figure 7 Two-dimensional intraoperative TEE midesophageal views (64° and 0°) with color flow Doppler demonstrating successful repair of the ruptured SVA. *RCC*, Right coronary cusp.

must be obtained to confirm the size and origin of the SVA and exclude other associated abnormalities (e.g., VSD).

For untreated ruptured SVAs, the median survival is approximately 3.9 years.[12] Length of survival is dependent on four clinical variables: (1) the speed at which the rupture develops, (2) the volume of blood flowing through the rupture, (3) the chamber into which the rupture opens, and (4) the presence or absence of a concomitant VSD.[5] Immediate death from a ruptured SVA is often caused by extravasation into the pericardium or into the base of the ventricular septum, which can lead to complete heart block and hemodynamic collapse. Although ruptured SVAs are usually managed surgically, a percutaneous closure may be a safe alternative in selected patients, but there are limited long-term outcome studies. Surgical repair of ruptured SVAs is mainly successful. Procedural mortality is estimated to be 1.9%-3.6%, with survival close to 95% at 20 years after surgery.[13]

## CONCLUSIONS

A ruptured SVA is a rare complication and requires early identification for prompt surgical intervention. This case underscores the importance of a thorough cardiac workup in young patients with nonspecific symptoms, unexplained dyspnea, and a diastolic murmur without any significant comorbidities. Echocardiography remains the modality of choice during the evaluation of and intervention with an SVA and provides real-time assistance in identifying subtle, yet important, findings that can alter the surgical plan. Early recognition and referral to surgery are important in achieving a good clinical outcome, but due to interstudy variability and the rarity of this echo finding, case reports such as this are exceedingly important to help inform the wider echocardiography community.

## SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2022.02.005.

## REFERENCES

1. Prian GW, Diethrich EB. Sinus of Valsalva abnormalities: a specific differentiation between aneurysms of and aneurysms involving the sinuses of Valsalva. Vasc Surg 1973;7:155-64.
2. Feldman DN, Gade CL, Roman MJ. Ruptured aneurysm of the right sinus of Valsalva associated with a ventricular septal defect and an anomalous coronary artery. Tex Heart Inst J 2005;32:555.
3. Arcos LC, Medina HM, Sandoval N, Gelves J, Salazar G. A case of sinus of Valsalva aneurysm rupture in a patient with bicuspid aortic valve. CASE (Phila) 2020;4:47-52.
4. Weinreich M, Yu PJ, Trost B. Sinus of Valsalva aneurysms: review of the literature and an update on management. Clin Cardiol 2015;38:185-9.
5. Takach TJ, Reul GJ, Duncan JM, Cooley DA, Livesay JJ, Ott DA, et al. Sinus of Valsalva aneurysm or fistula: management and outcome. Ann Thorac Surg 1999;68:1573-7.
6. Kalil R, Spitz J, Sciria C, Kim J, Narula N, Karas M, et al. Ruptured sinus of Valsalva aneurysm: an unusual cause of heart failure in a young woman. CASE (Phila) 2021;6:27-30.
7. Sengodan P, Kharazi A, Akhter SA, Nekkanti R. Rare presentation of a ruptured sinus of Valsalva aneurysm. CASE (Phila) 2020;4:43-6.
8. Jain PK, Narula J, Hasija S, Kiran U. Is it really ruptured sinus of Valsalva? The crucial role of comprehensive transesophageal echocardiography in clinical decision-making. Ann Cardiac Anaesth 2015;18:221-4.
9. Togashi K, Paez FJG, Sheu RD. Sinus of Valsalva aneurysm rupture associated with a ventricular septal defect: the importance of multi-angle assessment by intraoperative transesophageal echocardiography. J Cardiothorac Vasc Anesth 2020;34:3376-81.
10. Cheng TO, Yang YL, Xie MX, Wang XF, Dong NG, Su W, et al. Echocardiographic diagnosis of sinus of Valsalva aneurysm: a 17-year (1995-2012) experience of 212 surgically treated patients from one single medical center in China. Int J Cardiol 2014;173:33-9.
11. Martín AG, Ruiz JMO, González AE, García JMM, Benito F, Daza JAS. Multiplane transesophageal echocardiography in the preoperative evaluation of the sinus of Valsalva fistula to right chambers. [article in Spanish]. Rev Esp Cardiol 2002;55:29-36.
12. Vural KM, Şener E, Taşdemir O, Bayazıt K. Approach to sinus of Valsalva aneurysms: a review of 53 cases. Eur J Cardio-Thorac Surg 2001;20:71-6.
13. Van Son JA, Danielson GK, Schaff HV, Orszulak TA, Edwards WD, Seward JB. Long-term outcome of surgical repair of ruptured sinus of Valsalva aneurysm. Circulation 1994;90(5 Pt 2):II20-9.