Successful Treatment of Ruptured Sinus of Valsalva Aneurysm Associated With Subarterial Ventricular Septal Defect

Hieu L. Nguyen, Associate Professor, MD, PhD, Ly T. M. Nguyen, MD, PhD, Duy Q. Tran, MD, Jeroan J. Allison, Professor, MD, MS, Hoa Nguyen, Associate Professor, MD, PhD, David D. McManus, MD, ScM, and Anh L. Nguyen, BSN, *Hanoi, Vietnam; and Worcester, Massachusetts*

INTRODUCTION

Sinus of Valsalva aneurysm (SOVA) is rare, accounting for approximately 0.1% to 3.5% of all congenital heart diseases.[1] Ruptured SOVA can be attributed to congenital or acquired conditions such as degenerative connective tissue diseases (Marfan syndrome and Ehlers-Danlos syndrome) and infections (infectious endocarditis and syphilis).[1] In this case report, we present the successful surgical repair of a 21-year-old patient in Vietnam who had a subarterial ventricular septal defect (VSD) complicated by a ruptured SOVA in the right ventricle.

CASE REPORT

A 21-year-old man was hospitalized with acute dyspnea and high fever. Medical history revealed a previously diagnosed asymptomatic subarterial VSD approximately 4 mm in diameter. When the patient was 11 years old, a routine checkup revealed that the right coronary cusp had prolapsed into the VSD, resulting in mild aortic regurgitation (AR). While surgical intervention was indicated to address this defect, the parents decided to pursue watchful waiting given their assessment that the child was asymptomatic. Subsequent follow-ups of the patient's VSD were intermittent.

At the time of hospital admission, the patient presented with acute dyspnea, fever, and cyanosis (pulse oximetry, 90%). Clinical examination revealed a loud, harsh murmur over the third intercostal space at the left sternal border, heart rate of 120 beats per minute, blood pressure of 100/60 mm Hg, and coarse crackles in both lung fields. Electrocardiography showed sinus tachycardia and left ventricular hypertrophy (Figure 1A).

Chest radiography revealed diffuse bilateral opacification, interstitial thickening, and cardiomegaly (Figure 1B). Transthoracic echocardiography (TTE) revealed a large subarterial VSD with a left-to-right shunt, a moderately dilated left ventricle chamber with a normal ejection fraction, and elevated right ventricular systolic pressure. Laboratory tests revealed that the patient had severe metabolic acidosis, leukocytosis (29 g/L), elevated procalcitonin levels (5.1 ng/mL), and high NT-proBNP levels (828 pg/mL; normal range, <125 pg/mL).

These findings suggest decompensated heart failure complicated by severe pneumonia and acute respiratory distress syndrome. The patient was treated in the intensive care unit for a week with mechanical ventilation, continuous venovenous hemofiltration, and extracorporeal membrane oxygenation. The lung infection improved, and the patient was transferred to the heart center.

At the heart center, thorough and repeated TTE demonstrated a ruptured SOVA just above the VSD. Parasternal long- and short-axis TTE showed an 11-mm tunnel-like communication between the aorta at the right coronary sinus of Valsalva and the right ventricular outflow tract, both in two-dimensional (2D) and color-flow Doppler in diastole (Figure 2, Figure 3, Video 1, Video 2). In the apical 5-chamber view TTE, a subarterial VSD measuring 4 mm was observed both in 2D and color-flow Doppler in systole (Figure 4, Video 3), and a left-to-right continuous shunt flow was detected on continuous-wave Doppler signal across the VSD and ruptured SOVA (Figure 5). No findings of endocarditis were observed on TTE or transesophageal echocardiography (TEE, Video 4). The left ventricular dimensions, systolic function, and right ventricular systolic pressure were unchanged from those in a prior echo study.

The patient recovered over the following 2 months; however, the heart failure symptoms were difficult to control medically. The patient was dyspneic on mild exertion, and there was moderate lower extremity edema. Kidney function was abnormal. Surgical repair was necessary to close the ruptured SOVA and subarterial VSD.

Two months after admission, the patient underwent a midsternal thoracotomy under general anesthesia to access the heart. Through the aortic root opening on the right coronary sinus of Valsalva, the preoperative heart lesion was confirmed to be a ruptured SOVA (Figure 6A, Video 5) with a thin, fibrous structure extending 3.0 cm into the right ventricular outflow tract. The VSD situated beneath the 2 great arteries (Figure 6B) was then directly repaired. Finally, the ruptured SOVA was fixed using an artificial patch.

Upon cardiac reperfusion, TEE confirmed closure of the VSD and the absence of a residual shunt in the right sinus of Valsalva. Mild AR was observed. The patient's postoperative condition improved

From the Hanoi Medical University Hospital, Hanoi Medical University, Hanoi, Vietnam (H.L.N., L.T.M.N.); Cardiovascular Center, Hanoi Medical University Hospital, Hanoi, Vietnam (D.Q.T.); Department of Population and Quantitative Health Sciences, UMass Chan Medical School, Worcester, Massachusetts (J.J.A.); Division of Epidemiology; Department of Population and Quantitative Health Sciences, UMass Chan Medical School, Worcester, Massachusetts (H.N.); Department of Medicine, University of Massachusetts Chan Medical School, and UMass Memorial Health, Worcester, Massachusetts (D.D.M); and Department of Nursing - Midwifery, Hanoi Medical University, Hanoi, Vietnam (A.L.N.).

Keywords: Subarterial ventricular septal defect, Ruptured sinus of Valsalva aneurysm, Echocardiography, Imaging

Correspondence: Ly T. M. Nguyen, MD, PhD, Cardiovascular Center, Hanoi Medical University Hospital, Hanoi Medical University, 1 Ton That Tung, Hanoi, Vietnam. (E-mail: *minhly.ng@gmai.com*).

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VIDEO HIGHLIGHTS

Video 1: Two-dimensional TTE, parasternal long-axis view without *(left)* and with *(right)* color-flow Doppler, demonstrates the ruptured SOVA with a left-to-right shunt from the aorta toward the right ventricle.

Video 2: Two-dimensional TTE, parasternal short-axis view without *(left)* and with *(right)* color-flow Doppler, demonstrates a ruptured SOVA with a left-to-right shunt from the aorta toward the right ventricle.

Video 3: Two-dimensional TTE, apical 5-chamber view without *(left)* and with *(right)* color-flow Doppler, demonstrates normal biventricular size and systolic function, a subarterial VSD (best seen in systole), and a SOVA rupture (best seen in diastole).

Video 4: Two-dimensional TEE, long-axis *(left)* and short-axis *(right)* biplane display with color-flow Doppler, demonstrates the SOVA rupture into the right ventricular outflow tract.

Video 5: Surgical video during transaortic resection demonstrates the windsock-shaped SOVA at the right coronary sinus (tip of forceps).

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significantly with no signs or symptoms of heart failure. The patient fully recovered and was discharged 1 month after cardiac surgery. Six months after discharge, the patient underwent regular follow-up visits and was noted to be progressing without concerns.

DISCUSSION

A SOVA is a relatively rare congenital or acquired heart defect resulting from weakness of the elastic lamina between the aortic media and annulus fibrosus. Most aneurysms arise from the right or noncoronary sinuses and tend to protrude and rupture into the right ventricle or right atrium. The most common anomalies associated with ruptured SOVA are VSDs (30%-50%).[2]

A VSD with a diameter of 4 mm or less is considered small. Muscular VSDs have the highest rate of spontaneous closure, ranging from 80% to 90%. Perimembranous VSDs have a spontaneous closure rate of approximately 50% by the age of 2.[3] However, subarterial VSDs are unlikely to close spontaneously. In subarterial VSDs, the hemodynamic effects of the left-to-right shunt beneath the aortic valve leaflet result in gradual aortic valve prolapse and AR. The right coronary cusp of the aortic valve commonly prolapses, resulting in deformation of the Valsalva sinus and potential aneurysm. Aneurysms can progress asymptomatically over time. Regular follow-up is essential to detect aortic valve prolapse and significant AR, indicating the need for surgical repair to prevent rupture complications.

After rupture of the SOVA, a new diastolic shunt flow occurs from the aorta to the cardiac chamber, most commonly leading to rupture into the right ventricle. This occurs alongside the systolic shunt flow that moves from the left ventricle to the right ventricle through the VSD. Systolic murmurs associated with uncomplicated VSDs can become continuous murmurs if the SOVA ruptures. The severity of heart failure depends on the size of the rupture, which can range from mild to severe. In our case, the timing of the SOVA rupture was unknown. The patient was hospitalized because of a lung infection. When the patient was first admitted to the hospital, the acoustic windows were limited, and the opportunity to diagnose SOVA rupture was missed. However, due to clinical suspicion, this was reevaluated when the patient's condition improved.

In a patient with a history of subarterial VSD who has been previously asymptomatic and presents in this state, comprehensive examination and imaging with a high suspicion of SOVA rupture is necessary. Additionally, prompt echocardiographic examination is necessary to provide a definitive diagnosis.[4,5] A SOVA should be evaluated in multiple 2D views to visualize the characteristic windsock-shaped anechoic region and using color-flow Doppler to exclude rupture into the right heart. Continuous-wave Doppler should also be performed to document the timing as this is helpful to discriminate between a phasic VSD and continuous SOVA rupture spectral Doppler display profile. Continuous left-to-right shunt flow can be observed instead of the systolic left-to-right shunt of uncomplicated VSD. A windsock lesion associated with a VSD complicated by a ruptured SOVA, as seen on echocardiography, should be distinguished from a perimembranous VSD with accessory tissue that allows for spontaneous closure of the defect. In this situation, prior

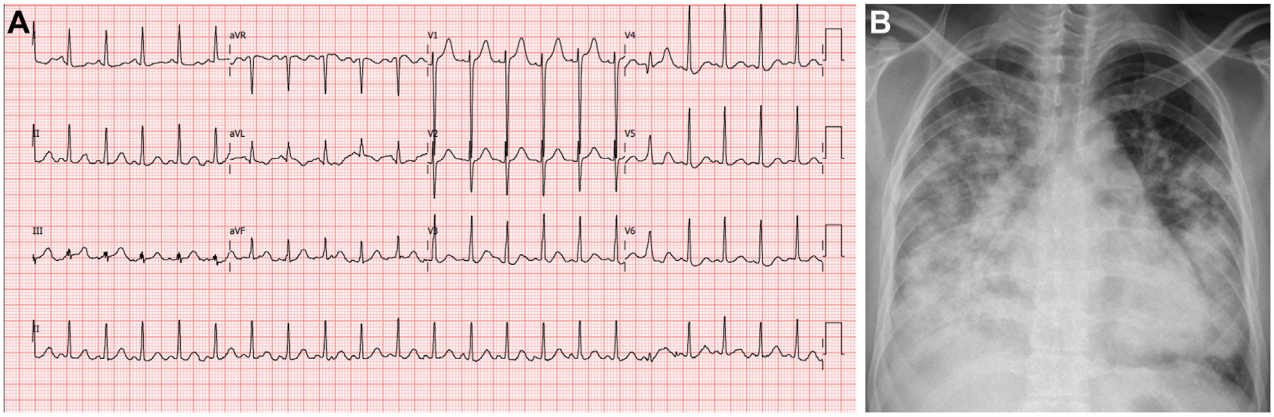

Figure 1 **(A)** Twelve-lead electrocardiogram demonstrates sinus tachycardia with voltage criteria for left ventricle hypertrophy; **(B)** chest radiography, posterior-anterior projection, demonstrates diffuse bilateral opacifications and cardiomegaly.

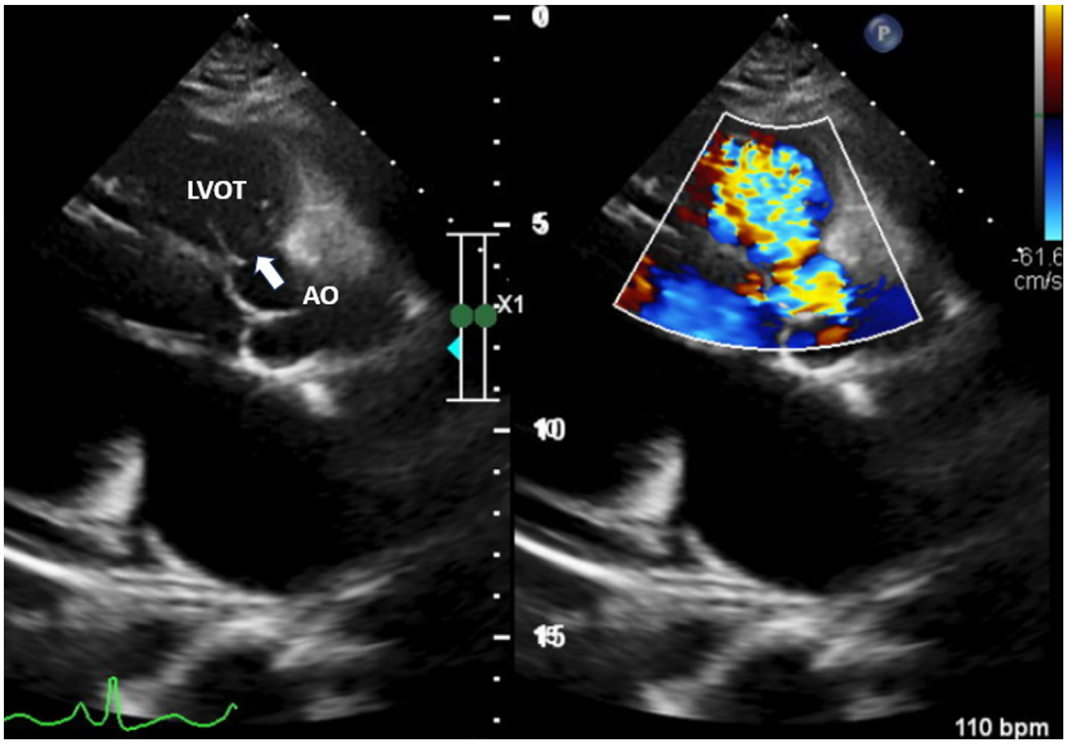

Figure 2 Two-dimensional TTE, parasternal long-axis diastolic view without *(left)* and with *(right)* color-flow Doppler, demonstrates a ruptured SOVA *(arrow)* with a left-to-right shunt from the aorta toward the right ventricle.

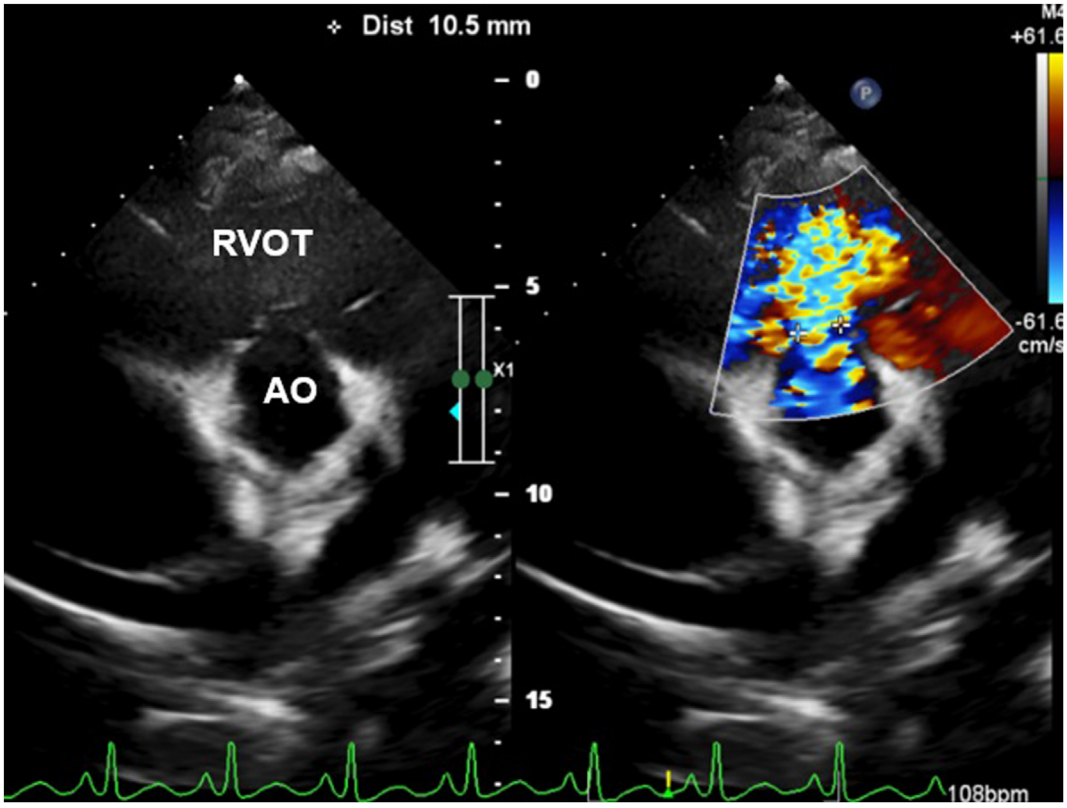

Figure 3 Two-dimensional TTE, parasternal short-axis diastolic view without (*left*) and with (*right*) color-flow Doppler, demonstrates a ruptured SOVA (10.5 mm) with a left-to-right shunt from the aorta toward the right ventricle.

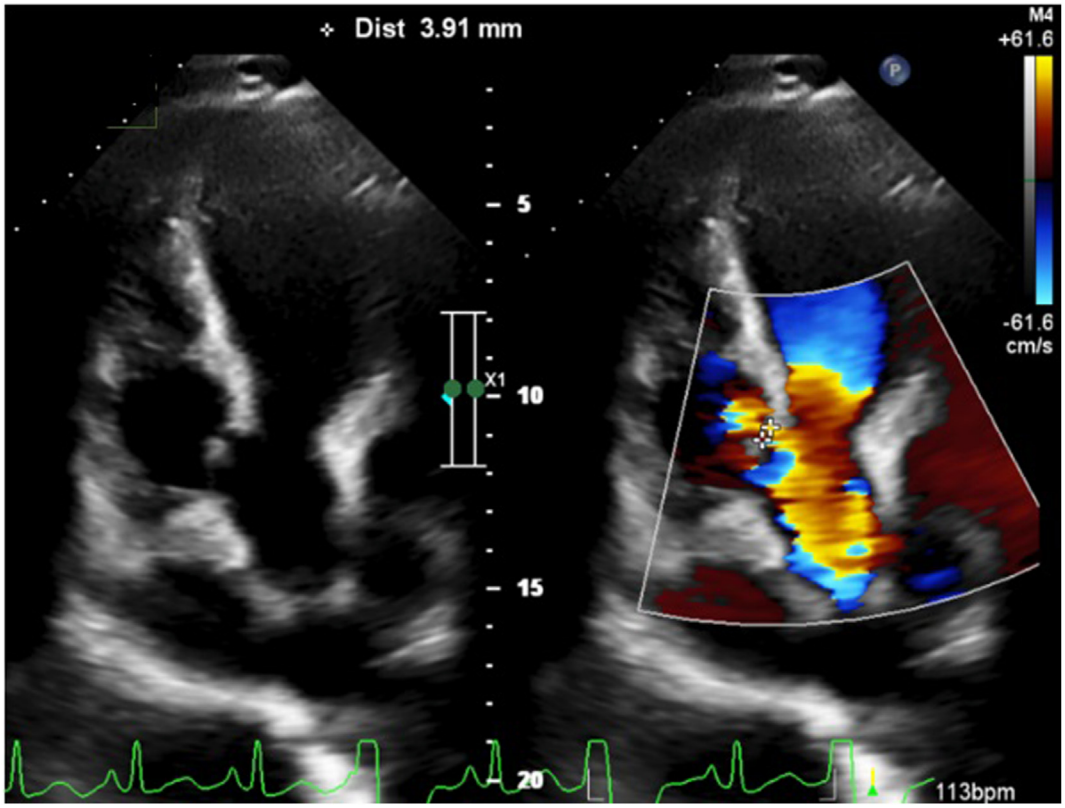

Figure 4 Two-dimensional TTE, apical 5-chamber systolic view without *(left)* and with *(right)* color-flow Doppler, demonstrates a subarterial VSD *(star)* with a left-to-right shunt *(cross)* from the left ventricle toward the right ventricle.

diagnoses and echocardiographic reports are important for an accurate assessment. Moreover, the variability in the lesion location noted in the parasternal short-axis view, along with the flow pattern across the defect, aids in making a definitive diagnosis.

In cases of suboptimal TTE images, TEE can be used to provide a more precise diagnosis. Both TTE and TEE are used to diagnose and monitor disease progression owing to their accessibility, flexibility, and reproducibility. In our case, TTE was performed several times and TEE was performed once during the course of treatment. After reviewing all the acquired images and loops, we discussed and agreed on the final diagnosis of subarterial VSD complicated by a ruptured SOVA. In our hospital, congenital heart disease and Doppler interrogation are beyond the scope of practice for most point-of-care ultrasonography users.

Cardiac computed tomography and cardiovascular magnetic resonance can provide enhanced imaging and detailed information about the coronary arteries and surrounding structures in cases of uncertain diagnoses.[6,7] In our case, we did not obtain any cardiac computed tomography or cardiovascular magnetic resonance images before surgery.

Surgical repair is the standard treatment for SOVA rupture.[8] The most commonly used surgical approach for treating ruptured SOVA is midsternal thoracotomy with cardiopulmonary bypass. A common "dual exposure" technique is used to access the aneurysm via the aortic root and the receiving chamber. This approach allows complete aneurysm removal, lesion closure, and evaluation and repair of the associated damage.[9] Depending on the location of the VSD, additional approaches are required to repair the entire lesion. The transaortic approach was chosen in our case because the VSD was located immediately below the aortic valve. Surgical repair of ruptured SOVA has a relatively low perioperative mortality rate. Moreover, the 10-year survival rate after surgery is 90%, and symptoms typically improve markedly.[10,11]

Device closure for cardiac aneurysms has emerged as a viable alternative to open-heart surgery. Because a device can be delivered and deployed via a catheter, sternotomy can be avoided and the patient can recover quickly. This treatment is typically indicated in patients with small-to-medium-sized ruptures of the SOVA without any associated anomalies, such as VSD or AR.[8,12,13]

CONCLUSION

Our case is one of several that presented with acute onset and severe symptoms but was ultimately cured.[14] A definitive diagnosis can be made through careful auscultation of the heart and thorough echocardiography. Considerable effort is required to treat a subarterial VSD complicated by a ruptured SOVA. We emphasize the importance of consistent follow-up with a cardiologist specializing in adult congenital heart disease to determine the appropriate timing for closing the defect rather than waiting for complications to arise.

ETHICS STATEMENT

The authors declare that the work described has been carried out in accordance with The Code of Ethics of the World Medical Association (Declaration of Helsinki) for experiments involving humans.

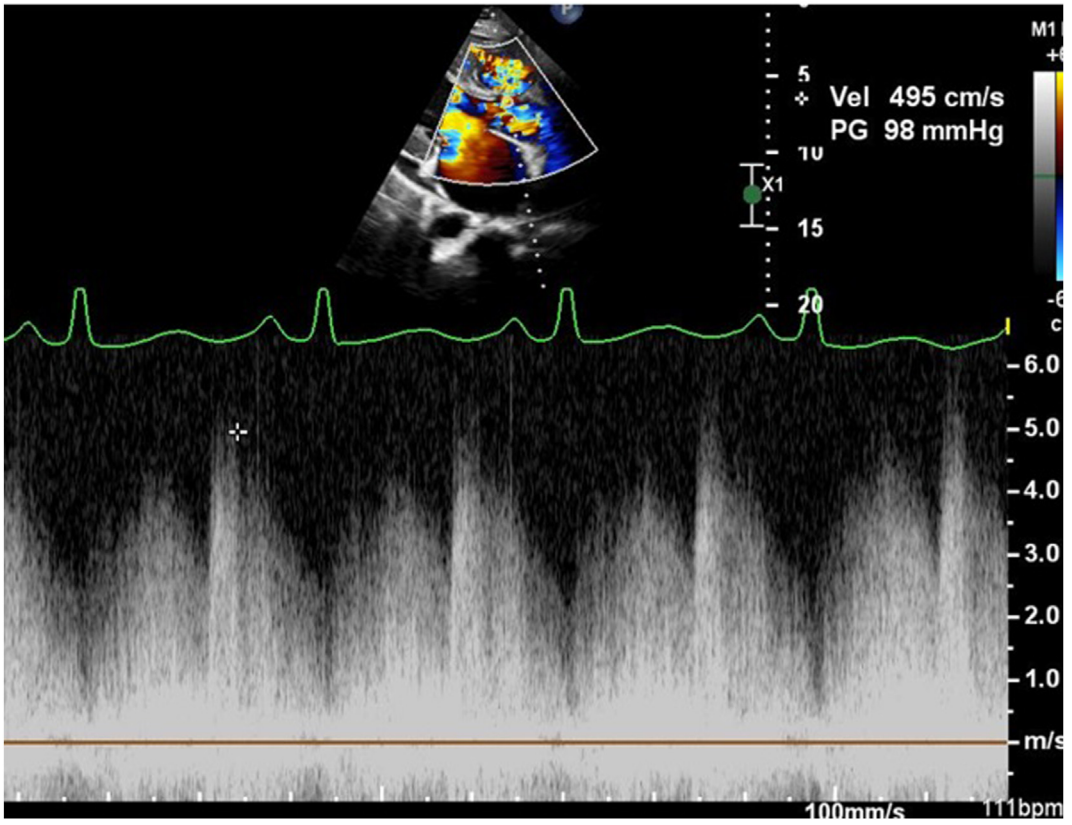

Figure 5 Two-dimensional TTE, parasternal long-axis view with color-flow Doppler-guided continuous-wave Doppler display, demonstrates the phasic flow across the VSD (higher velocity in systole) and the continuous flow from the ruptured SOVA (with the high diastolic flow velocity).

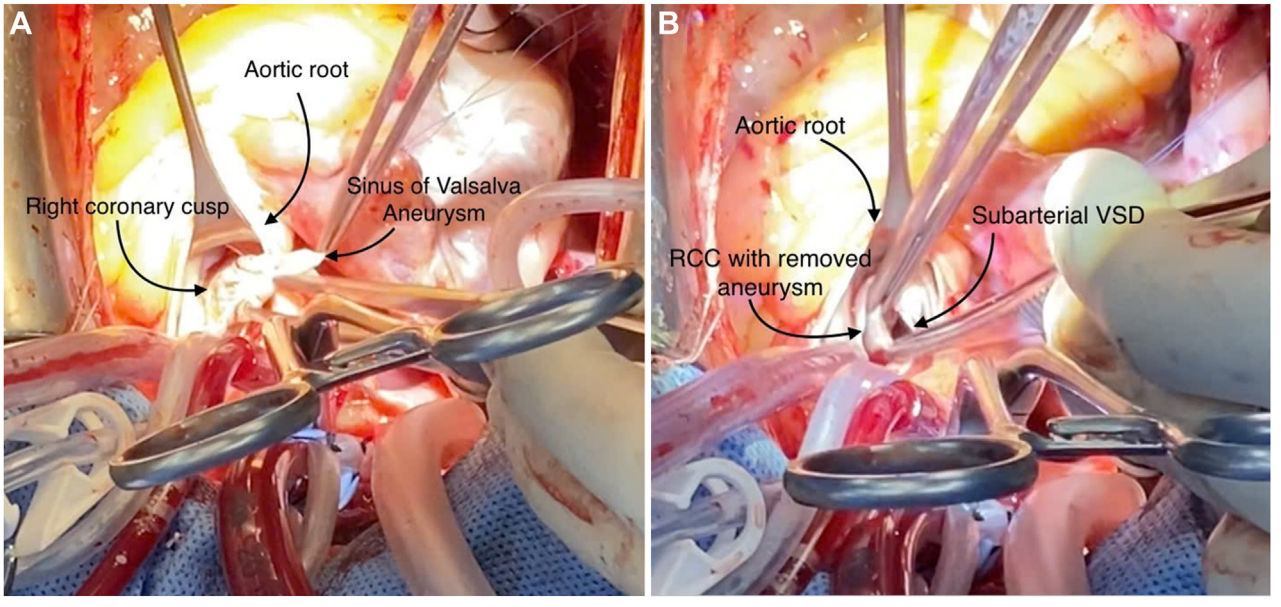

Figure 6 Surgical photograph of **(A)** the ruptured SOVA tissue (tip of forceps) and **(B)** the subarterial VSD exposed after resection of the SOVA. *RCC*, Right coronary cusp.

CONSENT STATEMENT

Complete written informed consent was obtained from the patient (or appropriate parent, guardian, or power of attorney) for the publication of this study and accompanying images.

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DISCLOSURE STATEMENT

The authors report no conflict of interest.

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SUPPLEMENTARY DATA

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