

TRICUSPID VALVE

CLINICAL CASE

Ruptured Sinus of Valsalva Aneurysm With Aorto-Atrial Fistula Mimicking Tricuspid Valve Endocarditis



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ABSTRACT

BACKGROUND Sinus of Valsalva aneurysm (SOVA) is a rare cardiac anomaly that can lead to acute cardiovascular collapse if ruptured. Prompt diagnosis and surgical repair are vital for survival.

CASE SUMMARY A 26-year-old woman presented with right upper quadrant pain and nausea. Work-up revealed elevated inflammatory markers, congestive hepatopathy, severe aortic regurgitation, and a suspected tricuspid valve vegetation. Infectious studies were negative. Transesophageal echocardiography ultimately identified a ruptured SOVA with an aorto-right atrial fistula. She underwent successful surgical repair and made a full recovery.

DISCUSSION Ruptured SOVA can mimic infective endocarditis and may be misdiagnosed without careful imaging. In this case, what was thought to be a vegetation was a ruptured aneurysm. Multimodality imaging played a key role in diagnosis, and early surgical repair was crucial.

TAKE-HOME MESSAGES Ruptured SOVA should be suspected in young patients with right heart failure and inconclusive infectious work-up. Early diagnosis ensures optimal outcomes. (JACC Case Rep. 2025;30:104561) © 2025 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

Sinus of Valsalva aneurysm (SOVA) represents a rare but potentially life-threatening cardiac anomaly characterized by dilatation of the aortic root between the aortic valve annulus and the sinotubular ridge. The prevalence of SOVA ranges from 0.09% to 0.5% of all cardiac disorders and constitutes up to 3.5% of congenital cardiac anomalies.¹ These aneurysms can be either congenital, associated with connective tissue disorders such as Marfan syndrome, or acquired secondary to infection, trauma, or degenerative changes.²

The clinical presentation of SOVA varies significantly, from asymptomatic incidental findings to acute cardiovascular collapse after rupture. Approximately 60% of cases involve the right ventricle, 30%

TAKE-HOME MESSAGES

- Ruptured SOVA should be suspected in young patients with right heart failure and inconclusive infectious work-up.
- Early diagnosis ensures optimal outcomes.

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**ABBREVIATIONS
AND ACRONYMS****SOVA** = sinus of Valsalva
aneurysm**TCC** = transcatheter closure**TEE** = transesophageal
echocardiography

involve the right atrium, and there is combined involvement of the left atrium, left ventricle, or pericardium in <10%.³ The anatomic distribution has significant clinical implications because rupture location determines the resulting hemodynamic compromise and clinical manifestations.

Rupture typically causes an abrupt left-to-right shunt with volume overload of the affected chamber, most commonly the right atrium/ventricle, leading to acute decompensated heart failure if left untreated.⁴

The diagnosis of ruptured SOVA can be challenging due to its rarity and clinical presentation that may mimic more common cardiovascular conditions such as infective endocarditis, especially when concurrent valvular abnormalities are present. Echocardiography remains the cornerstone of diagnosis, with transesophageal echocardiography (TEE) offering superior resolution to delineate the anatomic details necessary for surgical planning.

Surgical intervention and transcatheter closure (TCC) are both definitive treatments for ruptured SOVA, with contemporary data demonstrating excellent outcomes for surgical closure. Long-term survival after surgical repair ranges from 90% to 94% at 10 to 15 years.⁵ Factors influencing surgical approach include the size and location of the aneurysm, the presence of associated cardiac anomalies, and the degree of valvular involvement.⁶

We present a diagnostically challenging case of a 26-year-old woman who presented with nonspecific abdominal symptoms initially interpreted as tricuspid valve endocarditis on imaging. Further evaluation revealed a ruptured SOVA with an aortatrial fistula causing acute right-sided heart failure. This case highlights the importance of maintaining a high index of suspicion for uncommon cardiac anomalies in young patients presenting with new-onset murmurs and heart failure symptoms, even when initial imaging suggests alternative diagnoses.

HISTORY OF PRESENTATION

A 26-year-old woman presented to our institution with symptoms of right upper quadrant abdominal pain and nausea for 3 days before presentation. Initial vitals were unremarkable except for sinus tachycardia at 107 beats/min. Physical examination demonstrated a grade III/VI systolic murmur, louder on inspiration, best heard at the left lower sternal border, and significant right upper quadrant tenderness.

PAST MEDICAL HISTORY

She had a past medical history of cervical myelopathy status post C1 to C3 posterior fusion and C1 laminectomy.

DIFFERENTIAL DIAGNOSIS

Differential diagnoses included infective endocarditis, acute valvular regurgitation, acute myocarditis, acute pericarditis, and acute decompensated heart failure. We also considered gastrointestinal pathology like gall stones.

INVESTIGATIONS

Laboratory evaluation revealed significant abnormalities: elevated white blood cell count (20 K/ μ L), elevated C-reactive protein (7 mg/dL), and abnormal liver enzymes (aspartate transaminase: 338 U/L, alanine transaminase: 373 U/L). She had an elevated brain natriuretic peptide at 336 pg/mL. Troponin was within normal limits. Initial electrocardiogram showed sinus tachycardia with no ST-T segment abnormalities and normal intervals. Blood cultures were negative at 48 hours. Computed tomography angiography of the chest was negative for a pulmonary embolism and showed reflux of contrast into the intrahepatic inferior vena cava and hepatic veins. Computed tomography angiography also demonstrated a congenital right-sided aortic arch with a left innominate artery that divided into the left common carotid artery and left subclavian artery. Abdominal ultrasound was consistent with congestive hepatopathy. Initial transthoracic echocardiogram showed preserved left ventricular function (65%), normal right ventricular systolic function, and moderate aortic regurgitation with an eccentrically directed jet. A tricuspid valve vegetation (0.8 $\times$ 0.6 cm) was visualized on the atrial side of the valve with associated moderate regurgitation. Right ventricular systolic pressure was elevated at 56 mm Hg. Given the suspicion of infective endocarditis, a broad infectious work-up was sent, including Karius testing, and multiple serologies, including *Bartonella*, *Coxiella*, *Brucella*, and *Legionella*, which were all negative.

MANAGEMENT

The patient was covered with parenteral antibiotics, and we obtained a TEE to evaluate the vegetation further, which confirmed a shaggy vegetation on the atrial side of the tricuspid valve septal leaflet (measuring 11 and 6 mm in its largest segments)

(Figure 1). Additionally, a 6-mm perforation was identified in the aortic root near the sinus of Valsalva at the commissures of the noncoronary and right coronary cusps (Figure 2). This perforation resulted in severe aortic regurgitation directed into the right atrium; however, the aortic diameter remained normal. These findings supported a diagnosis of tricuspid valve endocarditis with a sinus of Valsalva perforation causing aortic regurgitation. An infectious diseases specialist was consulted and recommended to hold off on antibiotics, given negative cultures.

She was evaluated by the cardiothoracic surgery, which led to a recommendation for operative repair of the sinus of Valsalva and possible tricuspid and aortic valve replacement. The decision to proceed with surgical repair was reached after a multidisciplinary team discussion including a cardiac surgeon and interventional cardiologist. This was driven by the patients' low surgical risk and initial suspicion of infective endocarditis and possible aortic or tricuspid valve repair. Our patient also underwent a diagnostic coronary angiogram, given the aforementioned computed tomography chest finding of congenital right-sided aortic arch, to rule out congenital coronary anomalies, which revealed angiographically normal coronary arteries. Under general anesthesia, median sternotomy was performed and she was subsequently placed on on-pump cardiopulmonary bypass; the patient was observed to have a ruptured aneurysm of the non-coronary sinus of Valsalva, with direct communication into the right atrium. This presented as a distinct windsock deformity (Figure 3) extending from the aorta into the right atrial cavity. Intraoperative examination revealed no macroscopic evidence of infection, and both the tricuspid and aortic valves appeared free of vegetations. Given the proximity of the fistulous opening to the aortic valve leaflet, the defect was addressed by placing sutures originating below the aortic annulus within the left ventricular outflow tract and exiting through the noncoronary sinus. The resulting defect was then closed with a bovine pericardial patch, and most of the windsock deformity was excised. Intraoperative TEE confirmed completed repair (Figure 4). Her postoperative recovery was uneventful, and she was discharged on the fifth postoperative day, continuing to do well at the time of this report.

DISCUSSION

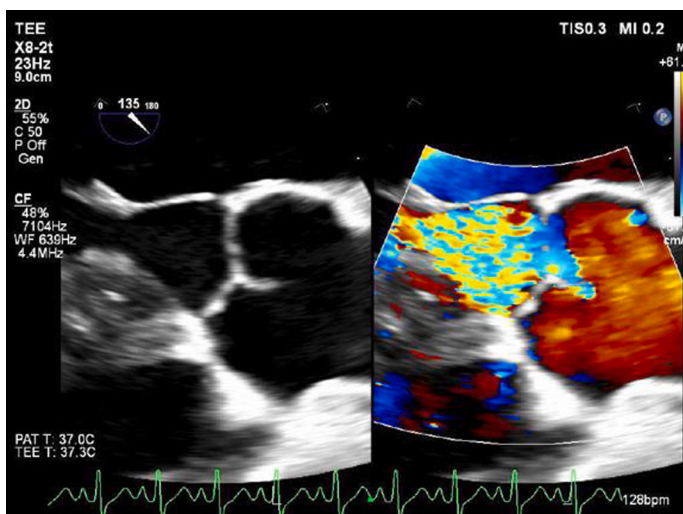
This case illustrates several important clinical pearls regarding the diagnosis and management of ruptured SOVA. First, the presentation of ruptured SOVA can

FIGURE 1 Preoperative Transesophageal Echocardiography Showing a Small, Shaggy Vegetation on the Atrial Side of the Tricuspid Valve Septal Leaflet



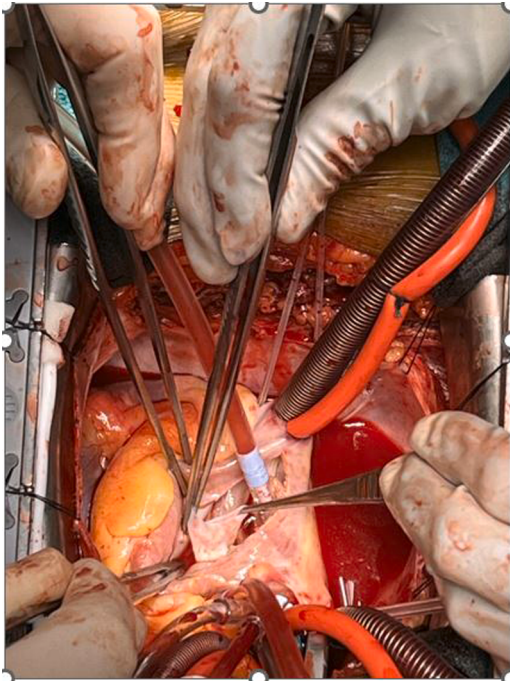
be highly variable and may mimic other cardiac conditions, particularly infective endocarditis. Our patient's presentation with right upper quadrant pain, elevated inflammatory markers, and an apparent vegetation on echocardiography led to an initial misdiagnosis of tricuspid valve endocarditis. However, the negative infectious work-up and intraoperative findings ultimately revealed a ruptured

FIGURE 2 Preoperative Transesophageal Echocardiography Showing 6-mm Perforation Into the Aortic Root Near the Sinus of Valsalva



This is near the commissures of the noncoronary cusp and right coronary cusp. Severe aortic regurgitation through the perforation is shown, directed into the right atrium.

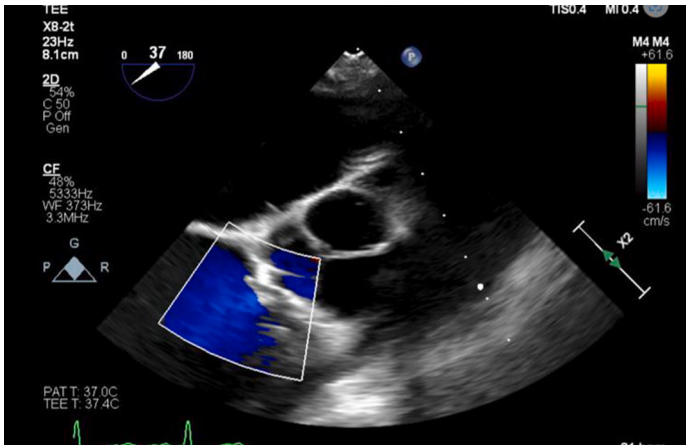
FIGURE 3 Intraoperative Image Showing Windsock Deformity



SOVA with aorto-right atrial fistula as the true etiology.

The pathophysiology of ruptured SOVA involves an abrupt left-to-right shunt with volume overload of the receiving chamber. In our case, rupture into the right atrium led to right atrial and ventricular volume overload, pulmonary hypertension, and congestive hepatopathy manifesting as right upper quadrant

FIGURE 4 Intraoperative Transesophageal Echocardiography Showing Completed Repair



VISUAL SUMMARY Summary of Events

Date	Events
Day 1 (date of submission)	A 26-y-old woman presented with right upper quadrant abdominal pain and nausea. Vitals notable for sinus tachycardia. Physical examination revealed a loud systolic murmur and right upper quadrant tenderness.
Day 2	Laboratory work-up showed leukocytosis, elevated C-reactive protein, liver enzyme derangements, and elevated brain natriuretic peptide. Abdominal ultrasound revealed congestive hepatopathy. Initial transthoracic echocardiography showed preserved left ventricular ejection fraction, moderate aortic regurgitation, and apparent tricuspid valve vegetation with moderate regurgitation.
Day 3	Infectious work-up was negative. TEE revealed a perforation in the noncoronary sinus of Valsalva with left-to-right shunting into the right atrium, mimicking tricuspid valve vegetation.
Day 4	Multidisciplinary team decision made for surgical intervention. Underwent operative repair: excision of ruptured SOVA with patch closure using bovine pericardium; native tricuspid and aortic valves preserved.
POD 1-4	Recovery was uneventful. Postoperative TEE confirmed successful repair with no residual shunt or valve dysfunction.
POD 5	Discharged home in stable condition.
30-d follow-up	Doing clinically well with no sequelae.

POD = postoperative day; SOVA = sinus of Valsalva aneurysm; TEE = transesophageal echocardiography.

pain and abnormal liver enzymes. This constellation of symptoms may be mistaken for primary hepatobiliary pathology or right-sided heart failure from other etiologies if a careful cardiovascular examination is not performed.

TEE remains the gold standard for the diagnosis of SOVA; however, interpretation challenges exist. In our case, the protrusion of the aneurysm into the right atrium was misinterpreted as a vegetation on the tricuspid valve. This diagnostic pitfall has been described previously in the literature, where the windsock deformity characteristic of ruptured SOVA can be mistaken for valvular vegetation, particularly when it prolapses through an atrio-ventricular valve.⁷ This underscores the importance of comprehensive multimodality imaging, including TEE and potentially cardiac magnetic resonance or computed tomography angiography in complex cases.⁸

The treatment of a ruptured SOVA is complex and requires a multidisciplinary team approach. Currently, 2 main therapeutic routes exist: TCC or surgical repair. Various surgical techniques have been described, including transaortic, chamber, and combined approaches.⁹ TCC devices including

septal occluder devices, ductal occluder, or Amplatzer vascular plugs have been used; in a recent systematic review by Aryan et al,¹⁰ the success rate was 95.6%. Patients may be considered for TCC if the rupture originates from the noncoronary sinus or right coronary sinus, the defect is <10 mm, the EuroSCORE II is >20% indicating a high surgical risk, and there are no associated cardiac anomalies.¹⁰ However, data on TCC-directed treatment remain limited, and the approach is relatively new compared with traditional surgical correction.¹⁰

In our patient, patch repair using bovine pericardium was successfully performed with preservation of both the native aortic and tricuspid valves. This approach aligns with contemporary surgical strategies that prioritize valve-sparing techniques whenever feasible, to avoid the long-term complications associated with prosthetic valves.⁹

The prognosis after surgical repair of ruptured SOVA is generally favorable, with reported 10-year survival rates of 90% to 94%.⁵ However, long-term follow-up is essential to monitor for potential complications, including recurrence, patch dehiscence, development of aortic regurgitation, or progression of associated cardiac anomalies.

This case also highlights the importance of considering congenital cardiac anomalies in young adults presenting with new cardiac symptoms, even

in the absence of previously diagnosed congenital heart disease. Although our patient had no known cardiac history, the intraoperative findings of a small, foreshortened aorta with high commissures may have been suggestive of an underlying congenital predisposition to SOVA formation.

CONCLUSIONS

Ruptured SOVA represents a rare but important diagnostic consideration in patients presenting with acute-onset murmurs and signs of heart failure. The diagnosis can be challenging, particularly when the clinical and echocardiographic findings mimic more common conditions such as infective endocarditis. A high index of suspicion, comprehensive multimodality imaging, and early surgical or TCC are essential for optimal outcomes in these complex cases.

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KEY WORDS aorta, pulmonary hypertension, tricuspid valve