

# Rare Presentation of a Ruptured Sinus of Valsalva Aneurysm



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## INTRODUCTION

Sinus of Valsalva (SOV) aneurysm occurs infrequently, with an incidence of 0.14% to 3.5% in patients undergoing open heart surgery. SOV aneurysm may be congenital or acquired in origin and may rupture into any of the cardiac chambers, usually the right-sided ones, to form an aorticocardiac fistula.<sup>1</sup> Clinically SOV aneurysm, can be classified as acquired or congenital and ruptured or unruptured, whereas anatomically it can be classified on the basis of the sinus of origin and the chamber of penetration.<sup>2</sup> We present a case of bacterial endocarditis complicated by ruptured SOV aneurysm in a patient who presented to the hospital with vague symptoms. The patient underwent successful patch repair and was doing well at 10-month follow-up.

## CASE PRESENTATION

A 29-year-old woman with a history of hypothyroidism and bicuspid aortic valve presented to the hospital with generalized fatigue. She had been treated for flu-like symptoms at an urgent care facility 1 week before hospitalization, with partial resolution. There were no symptoms of chest pain, shortness of breath at rest or exertion, leg swelling, weight gain, palpitations, dizziness, presyncope, or syncope. She had not undergone any recent dental procedures and had no history of intravenous drug use. Physical examination revealed a loud, continuous murmur in the left and right parasternal areas from the second to fourth intercostal spaces. Blood cultures grew *Staphylococcus aureus*, and a third-generation cephalosporin was initiated.

Two-dimensional transthoracic echocardiography (TTE) showed a 1 × 1 cm echogenic structure (Figure 1) on the atrial aspect of the septal leaflet of the tricuspid valve. Other cardiac valves did not demonstrate definitive vegetations. The short-axis view on TTE at the base showed severe eccentric tricuspid regurgitation (TR; Figure 2), and the right ventricular inflow view (Figure 3, Video 1) confirmed severe TR, but the inferior vena cava was collapsible. A perforation of the tricuspid valve could not be ruled out at this time. The eccentric TR jet was well demonstrated in the subcostal view on TTE (Video 2). Subsequently, transesophageal echocardiography (TEE), performed the next day, did not reveal definitive evidence of

valvular endocarditis but instead showed a ruptured SOV aneurysm, contrary to the findings on TTE. The ruptured SOV aneurysm was saccular and thickened in appearance (Video 3). Color flow Doppler imaging showing a fistula that communicated with the right atrium during systole, with the flow directed toward the right ventricle in diastole, arguing against a true TR jet (Video 4). Color M-mode imaging showed evidence of the jet trajectory during systole and diastole (Figure 4). The modified midesophageal bicaval transesophageal view showed the tricuspid valve with an echogenicity that appeared to be a mass situated on the valve, which was the ruptured SOV aneurysm (Video 5, Figure 5).

During surgery, the fistula was visualized at the noncoronary sinus and was approximately 1.5 cm in size (Figure 6). A pericardial patch was fashioned to repair the defect without creating any tension on the aortic annulus (Figure 7). A congenital bicuspid aortic valve with two fully developed commissures and a raphe between the right and left coronary cusps with no stenosis or regurgitation was seen intraoperatively (Sievers type 1, left-right fusion). No vegetations were identified on the tricuspid valve, but there was evidence of thickening surrounding the ruptured fistula. Despite using a sufficiently sized bovine pericardial patch for the repair without any tension, there was mild distortion of the annulus, which led to grade III aortic insufficiency (Figure 8); subsequently an aortic valve replacement was performed with a bioprosthetic valve. Cultures of the ruptured aneurysm were positive for *S aureus*; thus prolonged antibiotics were required for a total duration of 6 weeks. Surgery was performed 2 weeks after the identification of positive blood cultures. The patient had an uneventful postoperative course and was doing well at 10-month follow-up.

## DISCUSSION

The discrepancy in findings between TTE and TEE makes the present case even more interesting. We present a case of ruptured SOV aneurysm presenting with bacterial endocarditis, which is not common. We believe that the SOV aneurysm was congenital and that the bacterial infection weakened the aneurysm, causing it to rupture. SOV aneurysm accounts for up to 3.5% of all congenital cardiac anomalies.<sup>3</sup> SOV aneurysms are most commonly congenital, due to an absence of continuity between the aortic media and the annulus fibrosis or a developmental structural defect in the aortic annulus itself, which can gradually give way under aortic pressure to form an aneurysm.<sup>4</sup> They can also be secondary to events and conditions that weaken the juncture between the media and the annulus fibrosus of the aorta, such as trauma, bacterial endocarditis, syphilis, cystic medial necrosis, and atherosclerosis.<sup>1</sup> SOV aneurysms originate predominantly from the right coronary sinus (70%) and are more prevalent in men and patients of Asian descent, commonly between 30 and 45 years of age.<sup>5</sup> Coexisting cardiac lesions are not uncommon, with the most common being ventricular septal defects and aortic regurgitation.<sup>3</sup> The

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## VIDEO HIGHLIGHTS

**Video 1:** TTE mimicking severe TR.

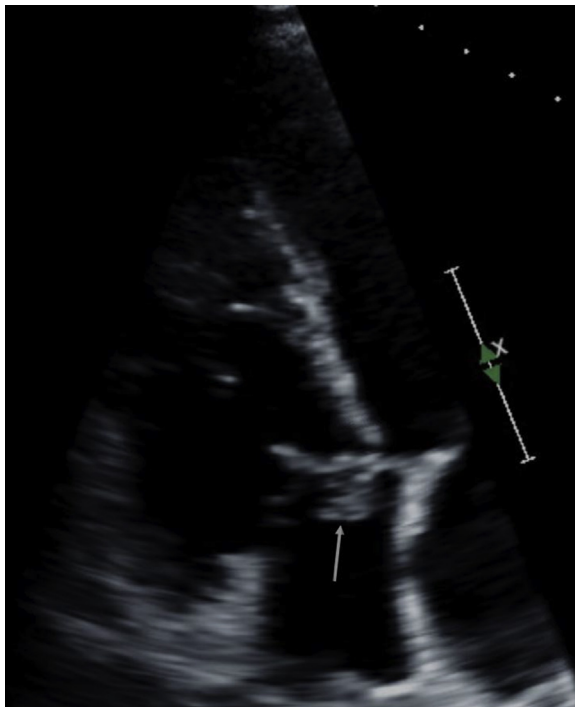
**Video 2:** Subcostal view on TTE showing eccentric TR.

**Video 3:** TEE, short-axis view at the base, showing ruptured SOV.

**Video 4:** TEE, short-axis view at the base, showing ruptured SOV with color Doppler flow.

**Video 5:** TEE with modified midesophageal bicaval view showing tricuspid valve with an echogenicity appearing to be a mass.

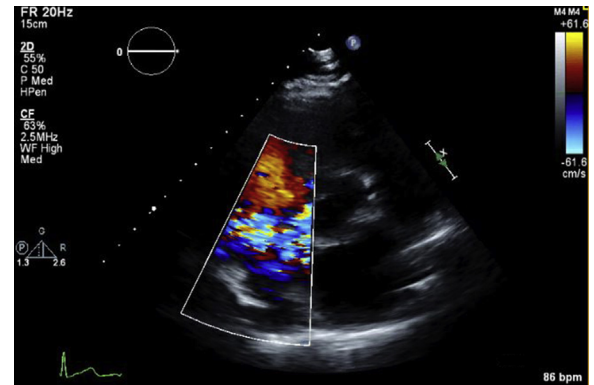
View the video content online at [www.cvcasejournal.com](http://www.cvcasejournal.com).



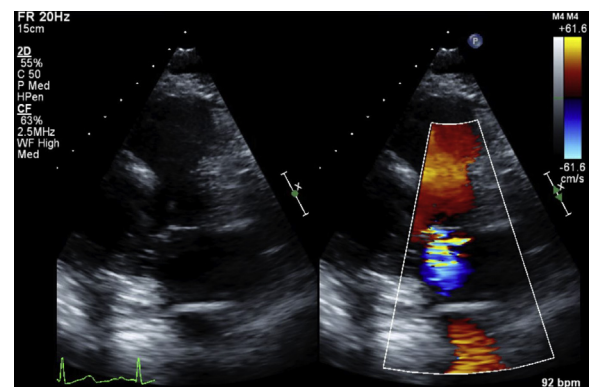
**Figure 1** Two-dimensional TTE showing a 1 × 1 cm mass on the atrial aspect of the septal leaflet of the tricuspid valve (arrow).

severity of symptoms depends on the size of the shunt, the presence of other cardiac lesions, and age at presentation.<sup>6</sup>

Bricker *et al.*<sup>7</sup> reported that rupture of an SOV aneurysm into the right ventricle was most common, with rupture into the right atrium being second most common, followed by the left ventricle and the left atrium. That can be attributed to the fact that SOV aneurysms usually affect the right coronary sinus, followed by the noncoronary sinus and finally the left coronary sinus. Patients with intact SOV aneurysms are usually asymptomatic, but once aneurysms have acutely ruptured, patients present with frank pulmonary edema as a result of sudden volume overload. Patients usually tolerate smaller, subacute perforations better as a result of progressive volume overload in which both ventricles dilate, thereby leading to decreased diastolic and systolic heart function over a period of time.<sup>8</sup> Very few



**Figure 2** TTE, parasternal short-axis view, mimicking severe TR.

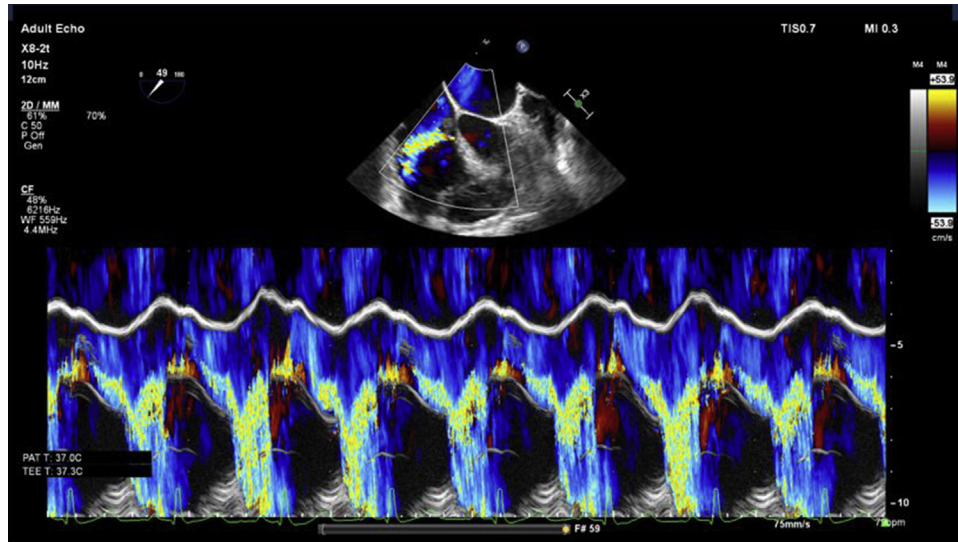


**Figure 3** TTE in the right ventricular inflow view consistent with severe eccentric TR.

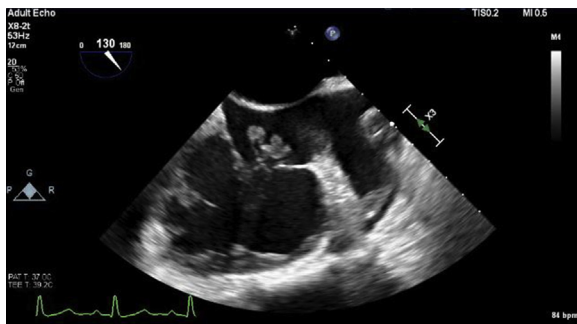
cases of asymptomatic ruptured SOV aneurysms similar to ours are described in the literature.<sup>8-11</sup>

Optimal imaging for SOV aneurysm is crucial, with the initial test of choice being TTE. Contrast echocardiography may be helpful in delineating the aneurysm and shunt arising from the rupture,<sup>12</sup> but TEE with color Doppler imaging is the test of choice for identifying rupture. The classic finding on TEE is a “wind-sock” extension of the sinus from body and/or apex of an otherwise normal aortic sinus when ruptured.<sup>2</sup> It is often helpful to perform multiplanar magnetic resonance imaging with sequences such as balanced steady-state free precession gradient-echo and half-Fourier acquisition single-shot turbo spin echo (“dark blood” imaging), which also allow accurate assessment of the origin and size of SOV aneurysms and the status of the surrounding cardiac and mediastinal anatomy and have the added advantage of quantifying the shunt and fistula flow.<sup>13</sup>

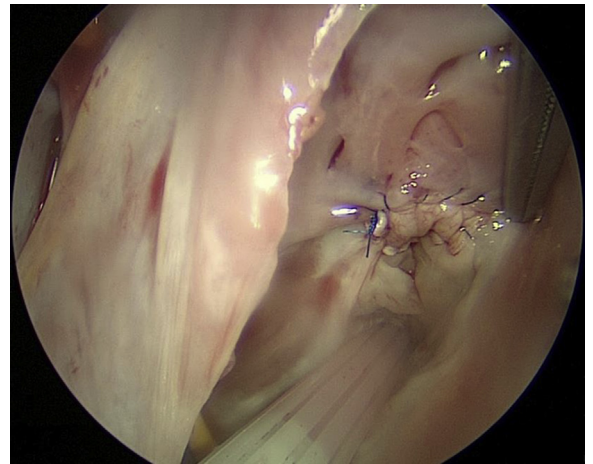
SOV aneurysms are typically surgically repaired when they rupture. The fistula may be closed with a pericardial or polyester patch or primarily with sutures, depending on the size of the defect. The management of an unruptured SOV aneurysm is not clear. Aortic regurgitation after surgical patch repair of the aneurysm is not uncommon<sup>3</sup> and has been reported similar to our patient. The risk of surgical treatment generally is low, with perioperative mortality rates ranging from 1.9% to 11.8% and 10-year survival rates ranging from 90% to 95%.<sup>10,13</sup> Our case highlights the fact that not all ruptured SOV aneurysms are symptomatic, and a high index of suspicion is needed when findings on TTE are inconclusive. In such cases, performing TEE without delay confirms the diagnosis.



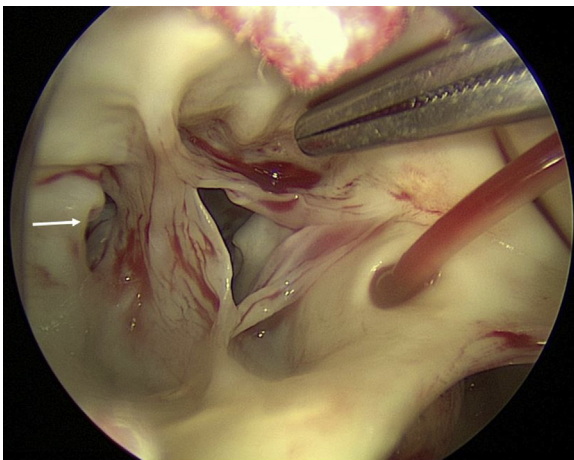
**Figure 4** Color M-mode imaging showing both systolic and diastolic flow.



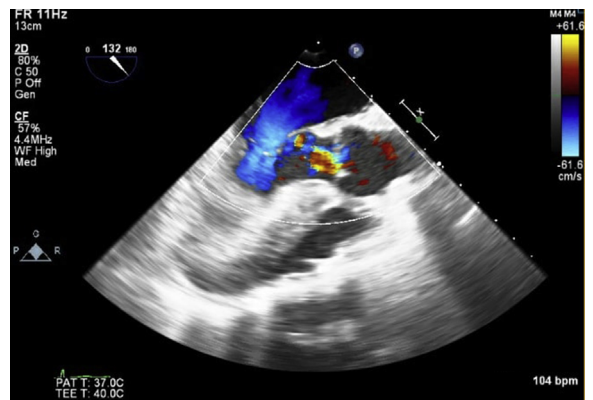
**Figure 5** Modified midesophageal bicaval view showing the echogenic structure, which appears to be on the tricuspid valve.



**Figure 7** Atrial side of the fistula visualized through the right atriotomy following repair with pericardial patch using running Prolene suture.



**Figure 6** A fistula (arrowhead) was visualized at the noncoronary sinus and was approximately 1.5 cm in diameter, as visualized through transverse aortotomy.



**Figure 8** TEE showing aortic insufficiency after patch repair of the aneurysm.

## CONCLUSION

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Rupture of a SOV aneurysm is usually accompanied by symptoms of acute dyspnea and heart failure. Nonspecific presentation of a ruptured SOV aneurysm complicated by bacterial endocarditis is rare. A ruptured SOV aneurysm traditionally requires surgical management, and prompt identification in the absence of classical symptoms is crucial for optimal outcomes.

## SUPPLEMENTARY DATA

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Supplementary data related to this article can be found at <https://doi.org/10.1016/j.case.2019.09.006>.

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