

# A challenging case report of acute sinus of Valsalva aneurysm rupture in cardiogenic shock and multi-organ failure with an emphasis on rapid recognition and ‘never giving up’ in face of futility

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## Background

Sinus of Valsalva aneurysm (SoVA) is a rare anomaly and can be divided into acquired and congenital forms, the latter being commonly associated with ventricular septal defects (VSDs). Rupture is a catastrophic complication with high mortality without urgent surgical intervention. We would like to highlight the use of echocardiography in an emergency setting for diagnosis and surgical intervention in a critically ill patient.

## Case summary

We report a 27-year-old female with history of conservatively managed VSD known since childhood. She presented with acute decompensated cardiac failure requiring intubation and inotropic support. Bedside echocardiography performed in the emergency department suggested a ruptured SoVA at the right coronary cusp with underlying supracristal VSD. Despite the patient being critically ill with multi-organ failure, surgery was performed as it was the patient's best chance for survival. Intraoperative findings tallied with the early echocardiographic results. She recovered gradually and was eventually discharged despite a stormy post-operative period.

## Discussion

This case report highlights the importance of prompt recognition of SoVA rupture by using bedside echocardiography. Surgical intervention needs to be early despite ongoing sepsis in view of acute mechanical failure. This case was unique as it illustrates a successful management of an acutely ill patient with multi-organ failure through early diagnosis, intensive perioperative stabilization, and surgical intervention.

## Keywords

Case report • Sinus of Valsalva aneurysm • Sinus of Valsalva rupture • Supracristal/doubly committed/subarterial ventricular septal defect

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## Learning points

- Sinus of Valsalva aneurysm is a rare cardiac anomaly and associated with various congenital heart defects mainly ventricular septal defects.
- Transoesophageal echocardiography remains the most indispensable tool in early diagnosis and in an emergency setting.
- Sinus of Valsalva aneurysm rupture needs to be addressed urgently with surgical intervention and sepsis should not delay surgery.
- Perioperative management of such cases can be very challenging.
- Surgical intervention should still be considered even in critically ill patients with multiple organ dama.

## Introduction

Sinus of Valsalva aneurysm (SoVA) is an aneurysmal dilatation of one or more aortic sinuses between the annulus of the aortic valve and the sinotubular junction. The congenital form is more common and frequently associated with ventricular septal defect (VSD).<sup>1-3</sup> Patients are generally asymptomatic until rupture occurs.<sup>3,4</sup> We present a case of SoVA rupture with underlying supracristal VSD and highlight the challenges in the diagnosis and management.

## Timeline

| Time  | Event   |
|-------|---|
| Day 0 | Started having malaise and productive cough   |
| Day 4 | Presented to an emergency department in a non-cardiac centre for sudden onset chest pain and dyspnoea.<br>Intubated and inotropic support initiated due to impending cardio-respiratory collapse.<br>Transthoracic echocardiogram showed a suspicious communication at the aortic valve area on PLAX and PSAX views.<br>Transoesophageal echocardiogram done immediately showed rupture of SoVA at the right coronary cusp with 2D colour depicting a turbulent jet from the aorta into the right ventricle.<br>First referral was made to the cardiothoracic surgeon. → plan for stabilization before transferring over.<br>Patient immediately transferred to intensive care unit for intensive care. |
| Day 5 | Patient deteriorated with worsening kidney and liver function.<br>Inotropic support increased.<br>Computed tomography thoracic and abdominal angiography performed urgently showed no evidence of aortic dissection.  |

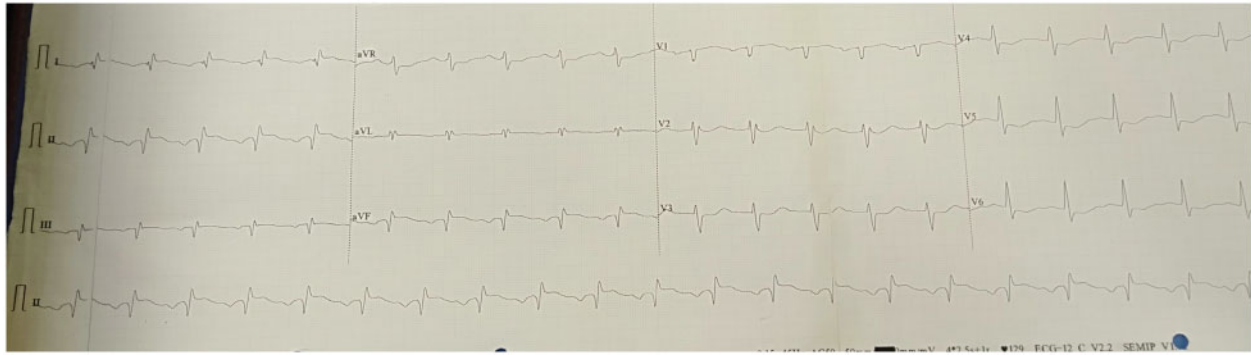
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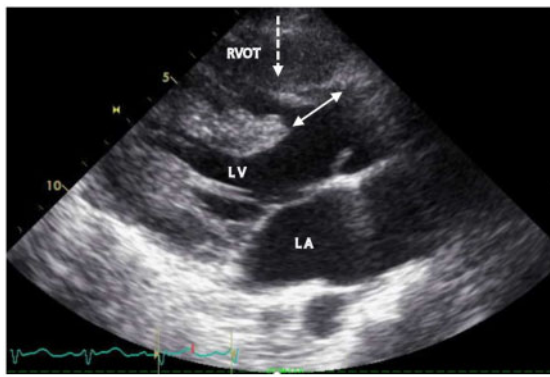
| Time   | Event  |
|--------|--|
|        | Referrals were attempted to other cardiothoracic surgeons within the area but unfortunately due to patient's worsening condition, patient was not stable for transfer.   |
| Day 6  | Referral was made to our national heart institute and she was accepted for transfer for emergency surgery.<br>Patient managed in cardiac care unit.<br>Placed on triple inotropic support and continuous veno-venous hemofiltration (CVVH)   |
| Day 7  | Cardiac arrest (PEA). Cardiopulmonary resuscitation for 7 min.<br>High-Risk Emergency Surgery [Repair of sinus of Valsalva aneurysm (SoVA) Rupture and ventricular septal defect Closure].   |
| Day 10 | Unstable AF (Cardioversion 3×).  |
| Day 12 | Patient regained consciousness as sedation was weaned off.   |
| Day 14 | Spiking temperature. Carbapenem and antifungal instituted.   |
| Day 15 | Cardiac Arrest (VF). External Defibrillation 4× (failed).<br>Chest was reopened urgently (internal defibrillation and cardiac massage).<br>No cardiac tamponade and no bleeding.<br>Pacing then initiated and subsequently blood pressure recordable.<br>Cardiopulmonary resuscitation was performed for 30 min. |
| Day 17 | CVVH stopped. Kidney and Liver function recovered.   |
| Day 18 | Chest closed (thoracotomy wound) surgically.   |
| Day 21 | Tracheostomy performed.  |
| Day 31 | Vital signs stable without inotropic support.  |
| Day 33 | Independent breathing without ventilation support.   |
| Day 34 | Started having intermittent fever. Consulted Infectious Disease team. Treated for mediastinitis. Cultures negative.  |
| Day 62 | Discharged well with regular follow-up and intensive rehabilitation programme for management of her critical illness polyneuropathy.   |

## Case presentation

A 27-year-old female presented acutely to an emergency department at a local hospital. Her symptoms began with malaise followed by productive cough, rhinorrhoea, and fever. She then had sudden onset of chest pain and dyspnoea. History from her caregiver indicated that she was known to have a cardiac murmur since she was 4 months old and had follow-up until 16, at which point she was discharged without surgical intervention. She did not have regular medication, history of tobacco nor illicit drug use. There was no recent travel history or close contact with persons with viral illness.



**Figure 1** Electrocardiogram at Emergency Department (1st encounter) showing suspicious changes for STEMI with ST-elevation in leads II, III, aVF with reciprocal changes in inferior leads (V4–V6).



**Figure 2** 2D Transthoracic echocardiogram. Parasternal long axis (PLAX) view showing defect in the ventricular septum measuring 1.9 cm (double-ended white arrow). Sinus of Valsalva aneurysm ruptured and prolapsed into the right ventricle (dotted arrow). LA, left atrium; LV, left ventricle; RVOT, right ventricle outflow tract.

On arrival, she was tachypnoeic with a respiratory rate of 24 breaths per minute and hypoxic requiring high flow mask oxygen of 15 L/min. She was also tachycardic at 130 b.p.m. and had a blood pressure (BP) of 125/53 mmHg. She was afebrile. Cardiovascular examination revealed a pansystolic murmur grade 2/6 heard best over the left upper sternal edge with chest auscultation revealing bilateral crepitations.

Her electrocardiogram showed ST-elevation in leads II, III, and aVF with ST depression in V4–V6 (Figure 1). Cardiac enzyme (Troponin I) was elevated at 6283 ng/mL. Her condition rapidly deteriorated requiring invasive ventilation. An urgent bedside transthoracic echocardiogram (TTE) showed normal left ventricle function without regional wall motion abnormality that would otherwise suggest a myocardial infarct. In parasternal long axis (PLAX) view, there was a defect in the ventricular septum denoting a VSD (Figure 2).

Interestingly, the parasternal short axis (PSAX) view revealed ruptured right SoVA with left-to-right shunt from the aorta into the right

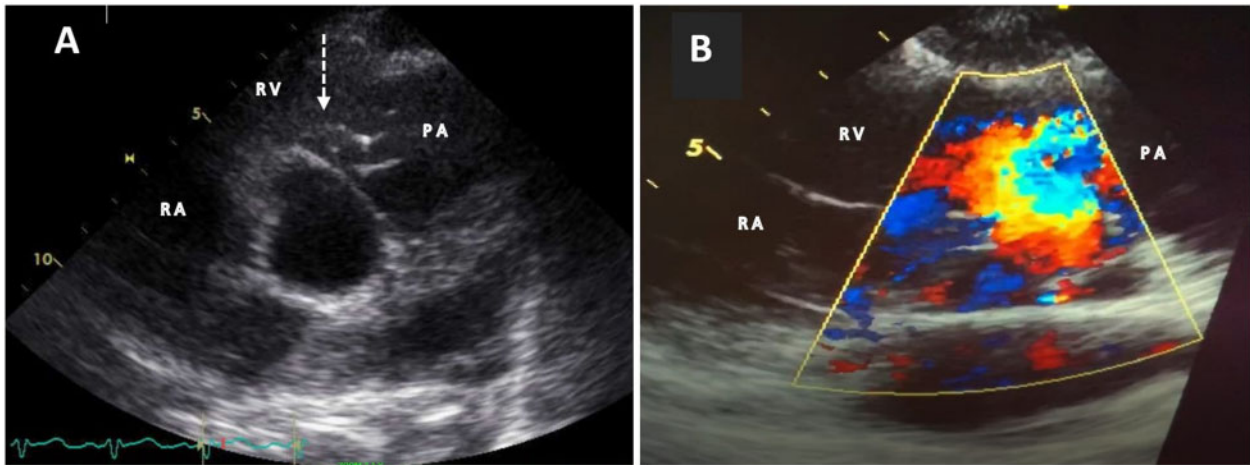
ventricular outflow tract (RVOT) near the pulmonic valve (Figure 3A and B). These findings refined our diagnosis to doubly committed subarterial VSD. This was further complicated by rupture of the SoVA prolapsing through the VSD into the RVOT giving rise to a 'windsock' appearance. There was also a suspicion of an intimal flap at the ascending aorta in the suprasternal view. Subsequently, a transoesophageal echocardiogram was performed (Figure 4) and confirmed the TTE findings.

An immediate referral was made to the nearest cardiothoracic centres for urgent surgical repair. Stabilization of patient's condition was emphasized before transferring over for surgery. The patient was admitted to the intensive care unit (ICU) and was treated empirically for infective endocarditis using local antibiotics guidelines.

During the first 2 days in ICU, the patient's condition continued to decline. She required double inotropic support with higher ventilator settings. The patient developed an acute kidney injury and acute liver failure. Her white cell count was  $31 \times 10^9/L$  (neutrophilia), urea and creatinine were 15.3 mmol/L and 321  $\mu\text{mol/L}$ , respectively. Her ALT (Alanine Transaminase) and AST (Aspartate Transaminase) were 3458 U/L and 5919 U/L, respectively.

Patient was finally referred to our centre for emergency surgery. Upon arrival in our cardiac care unit, she was critically ill needing multiple inotropes with overwhelming sepsis and disseminated intravascular coagulation. Haemodialysis was initiated in view of worsening kidney failure. Antibiotics were escalated to intravenous Piperacillin/Tazobactam. At one point, the patient went into cardiac arrest with pulseless electrical activity. Cardiopulmonary resuscitation was performed for 7 min before return of spontaneous circulation. Despite her dire prognosis, the multidisciplinary team decided to proceed with high-risk emergency surgery especially since she was of young age. Surgery revealed a large SoVA at the right coronary cusp with windsock prolapsing through a large (20 mm) subarterial VSD. The rupture was at the tip of the windsock (Figure 5). The surgeon closed the defects with GORE-TEX® patches.

On Day 3 post-surgery (POD 3), she had unstable atrial fibrillation needing electrical cardioversion. On POD 8, she had incessant ventricular fibrillation which did not respond to external defibrillation, necessitating the surgical team to perform emergency exploratory sternotomy for resuscitation. No cardiac tamponade and bleeding



**Figure 3** Ruptured Right sinus of Valsalva aneurysms on 2D transthoracic echocardiogram. Parasternal short-axis view of aortic valve and right ventricular outflow tract demonstrating (A). ‘Windssock’ appearance of ruptured sinus of Valsalva aneurysms as it prolapses into the right ventricular outflow tract near the pulmonic valve as shown by the dotted white arrow (B). Colour Doppler in systole showing distinct left-to-right shunt from the aorta into the RVOT. PSAX, parasternal short axis; RVOT, right ventricular outflow tract.

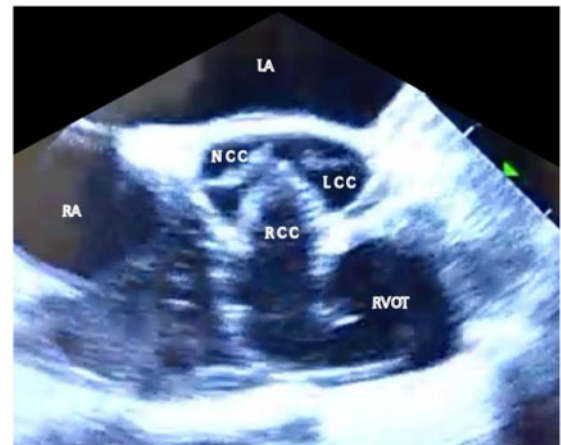
were found, and the patient responded to internal defibrillation and cardiac massage. Cardiopulmonary resuscitation was performed for 30 min followed with internal cardiac pacing. Resuscitation was successful and she gradually recovered. Antibiotics were also extended to a duration of 1 month for mediastinitis despite negative blood cultures.

After 6 weeks of hospitalization, she was discharged in good condition. She required intensive rehabilitation due to critical illness polyneuropathy.

## Discussion

Sinus of Valsalva aneurysms comprise 0.1–3.5% of all congenital heart defects with a male to female ratio of 4:1.<sup>2,3</sup> The prevalence of SoVA is also higher amongst the Asian population.<sup>2</sup> Often, patients with SoVAs are asymptomatic and the majority present with varied symptoms like cough, fatigue, chest pain, and dyspnoea.<sup>5</sup> Rupture often occurs frequently in the 3rd decade of life.<sup>5</sup> Our patient presented in such a manner and was diagnosed with echocardiography.

Her past history was the clue pointing towards an underlying congenital heart disease. The frequency of rupture SoVA varies according to location: 60% in right coronary sinus, 42% in non-coronary sinus, and only 10% in left coronary sinus.<sup>6</sup> Rupture can occur as a spontaneous event or due to trauma, extreme physical exercise, or infective endocarditis.<sup>6</sup> Supracristal VSD (SCVSD), also known as doubly committed subarterial, conal, or type 1 VSD accounts for 5–7% of all VSD.<sup>7</sup> In essence, the SCVSD produces a left-to-right shunt with a venturi effect that may eventually lead to herniation of the right aortic sinus with subsequent development of aneurysm and hence an acquired aortic valve deformity.<sup>8</sup> This contrasts to the congenital type of association between SCVSD and right coronary Valsalva sinus aneurysms. This is thought to relate to incomplete fusion of the

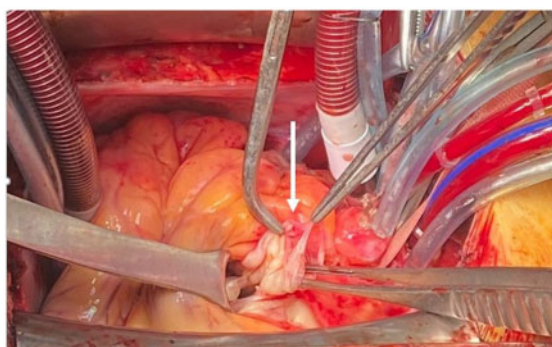


**Figure 4** Transoesophageal echocardiogram (midoesophageal parasternal short-axis view) showing classical ‘windssock’ appearance of ruptured right coronary sinus. LA, left atrium; LCC, left coronary cusp; NCC, non-coronary cusp; RA, right atrium; RCC, right coronary cusp; RVOT, right ventricular outflow tract.

truncal swellings at the time of the division of the common truncus from the bulbar septum during the 5th week of embryogenesis.<sup>8</sup>

In our patient, although no vegetation was detected by echocardiography, the ruptured SoVA may have been triggered by an infective process which weakened the aneurysm leading to rupture. An acute perforation at a high flow area would not allow for haemodynamic compensation. This may be the reason why our patient deteriorated rapidly.<sup>9</sup> Patients may also present with chest pain mimicking acute myocardial infarct<sup>10</sup> such as in our case. Echocardiography remains indispensable as it aids in detecting functional and structural





**Figure 5** Intraoperative finding showing right coronary sinus of Valsalva aneurysm rupture.

abnormality given that our patient did not have traditional risk factors for ischaemic heart disease.

Sinus of Valsalva aneurysms are viewed best as depicted in [Figures 2 and 3A and B](#). Computed tomography and magnetic resonance imaging could be used but may not be suitable in emergency setting.<sup>3,9</sup> Even with the advent of transcatheter closure devices for non-ruptured SoVA, surgical closure remains the definite treatment for ruptured SoVA with an operative mortality rate as high as 3.6%.<sup>11,12</sup> In settings of unruptured SoVA, surgery is recommended for those associated with VSD, significant aortic regurgitation, symptomatic, or enlarging SoVA.<sup>12</sup> At long-term follow-up after surgery, it is recommended to assess patients clinically such as by NYHA status and regular echocardiograms to look for recurrence of SoVA, previous valvular defects, and ventricular function.<sup>13</sup> To the best of our knowledge and based on current literature review, there are no reports of survivors with or without surgery in such a clinical scenario. Therefore, there is a role for surgery in a young patient with multi-organ failure who has undergone cardiopulmonary resuscitation.

## Lead author biography



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'Young Investigator Award' in the Malaysian Society of Hypertension Annual Scientific Meeting in 2019.

## Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

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**Slide sets:** A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

**Consent:** The authors confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

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