

# Multilobulated Sinus of Valsalva Aneurysm Dissecting into the Interventricular Septum (DAIS) and Rupturing into Left Ventricle: A Case Report

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## Background

Dissecting aneurysm of sinus of Valsalva (SOV) into the interventricular septum is a rare entity. Multilobulated form of dissection rupturing into the left ventricle (LV) has never been reported in the literature.

## Case summary

A 52-year-old male presented with dyspnoea and palpitation with wide pulse pressure and peripheral signs of distal run-off and a continuous murmur along the left parasternal area. Echocardiography revealed dilated right coronary cusp (RCC), which burrowed into the interventricular septum (IVS), forming multi-loculated cystic lesion which ruptured into LV with associated restrictive ventricular septal defect (VSD) and severe aortic regurgitation. Computed tomography (CT) angiography confirmed a 4.8 cm × 5.3 cm × 5.4 cm multiseptated aneurysm. The surgery involved excision of the aortic valve (AV) with the sinus, ligation of its penetrating portion at the crest of IVS, closure of VSD, and AV replacement. Postoperative echocardiography showed the complete collapse of the IVS component of the SOV aneurysm and the normally functioning mechanical AV.

## Discussion

Dissecting aneurysm into the IVS is a rare variant of SOV aneurysm, usually arising from RCC. It is mostly congenital in origin and has wide variety of presentations like congestive heart failure, palpitations, recurrent syncope, chest pain, sudden cardiac arrest, infective endocarditis, cerebral infarction, or asymptomatic. Aortic regurgitation is present in 30–50% of cases. Conduction disturbances from first-degree block to complete heart block are common. Echocardiogram, CT angio, and magnetic resonance imaging are useful for diagnosis. Surgical repair is the only option for treatment.

## Keywords

Sinus of Valsalva aneurysm • Dissecting aneurysm of the interventricular septum (DAIS) • Rupture of the sinus of Valsalva • Echocardiography • Computed tomography (CT) angiography

## Learning points

- Dissecting aneurysm into the interventricular septum (DAIS) is a rare variant of sinus of Valsalva aneurysm. Usually arises from right coronary cusp due to its proximity to interventricular septum.
- CHF, palpitations, recurrent syncope, chest pain, sudden cardiac arrest, infective endocarditis, and cerebral infarction are different kind of presentation. Aortic regurgitation in 30–50% of cases.
- Conduction disturbances common: first-degree block to complete heart block.
- Diagnosis with echocardiogram, computed tomography, and cardiac magnetic resonance imaging.
- Surgical repair is the only option. Perioperative death rate 3.9%.

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## Introduction

Aneurysm of sinus of Valsalva (SOV) is an abnormal dilatation of the aortic root between the aortic valve (AV) annulus and the sinotubular junction, most commonly involving the right coronary sinus (RCC).<sup>1</sup> They mostly remain asymptomatic and detected incidentally. When ruptured, they manifest with heart failure. Rupture of SOV aneurysm (RSOV) is rare, and their dissecting aneurysm into the interventricular septum (DAIS) is scarcely reported.<sup>2–8</sup> In addition, a multilobulated form of dissection has never been described in the literature. Here, we present a rare case of RSOV, which had a multilobulated configuration and burrowed into the interventricular septum (IVS) to rupture into the left ventricle (LV).

## Timeline

Day 1	Patient presented to the outpatient department
Day 2	Patient underwent transthoracic echocardiography (TTE) [TTE showed dissecting aneurysm into the interventricular septum (DAIS) and aortic regurgitation]
Day 4	Computed tomography angiography confirmed DIAS
Day 8	Aortic valve replacement and repair of sinus of Valsalva aneurysm done
Day 15	The patient was discharged
Day 149	Most recent follow-up

## Case presentation

A 52-year-old gentleman presented with gradually progressive dyspnoea and palpitation for the last 6 months. There was no history of chest pain, syncope, presyncope, cough, or wheeze. There was no history of rheumatic fever or trauma or family history of Marfan's syndrome. On examination, the patient was tall, average-built, had high volume and collapsing pulse with a pulse rate of 104/min and blood pressure of 140/60 mmHg. Jugular venous pressure was elevated with prominent X and Y descent. Signs of distal run-off were present, i.e. Corrigan's pulse in carotids and pistol shot sounds over both femoral arteries. The apex was hyperdynamic and displaced laterally. He had prominent S3 and a to and fro murmur heard in the right third intercostal space and along the left parasternal area with a high-pitched ejection systolic murmur followed by a high-pitched blowing holo-diastolic murmur.

The electrocardiogram showed normal sinus rhythm with bifascicular block (Figure 1). An echocardiogram (Figure 2A–C; Video 1) revealed a multilobulated cystic lesion in the IVS. Colour-Doppler imaging revealed blood flow into these cystic lesions, expanding in diastole and decreasing in size during systole. Right coronary cusp was dilated and was found to burrow into the IVS leading to these cystic lesions. This lesion opened onto the left ventricular side of the septum, making a likely diagnosis of ruptured RSOV to LV after forming a multilobulated cystic course into the IVS. We could appreciate an associated ventricular septal defect (VSD) at the upper end of the

IVS, having bidirectional flow. The left ventricular ejection fraction (LVEF) was decreased (35%) with global hypokinesia. There was severe aortic regurgitation (AR) with holo-diastolic reversal in the descending thoracic aorta.

Computed tomography (CT) angiography (Figure 3) confirmed echocardiographic findings and showed a large multiseptated aneurysm with dimensions of 4.8 cm × 5.3 cm × 5.4 cm arising from RCC which was seen to burrow into the basal and mid-cavity IVS with mass effect on left ventricular outflow and communicating with LV.

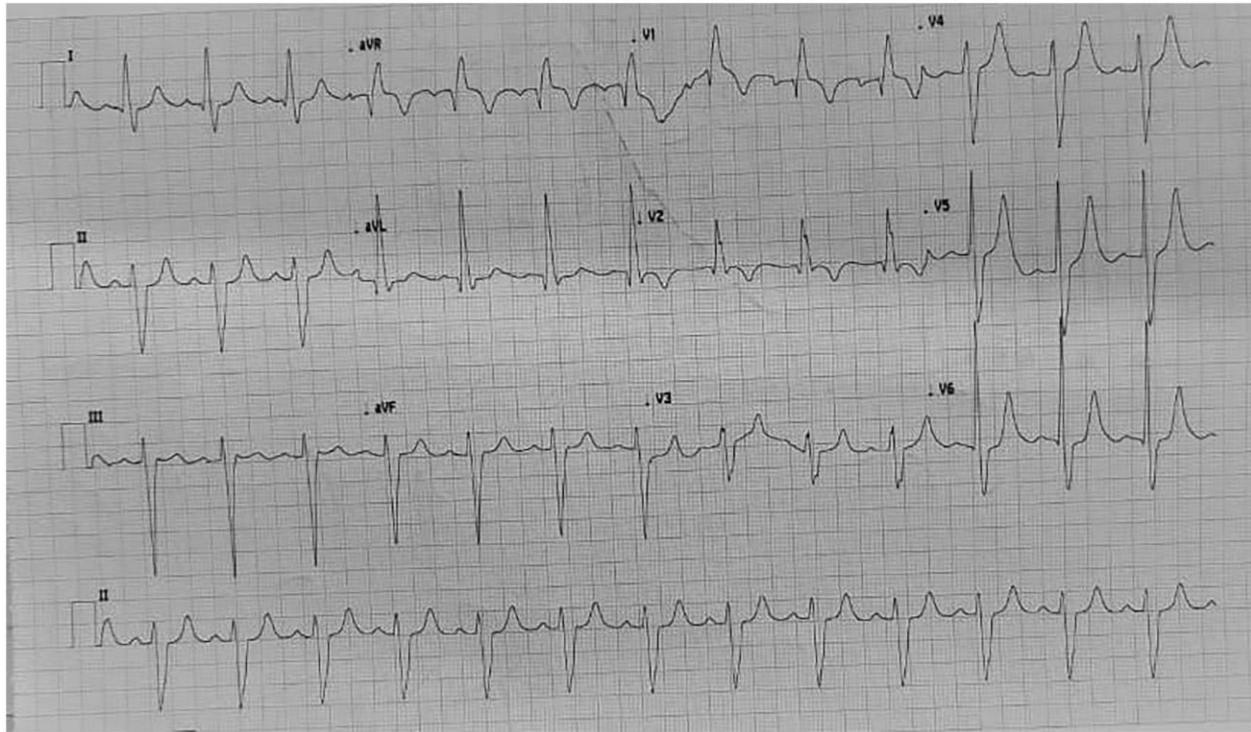
After discussion with the heart team, he was taken up for surgical repair of RSOV. Preoperative coronary angiography with aortic root angiogram showed right dominant normal coronaries with RSOV through IVS to LV. Intraoperatively, AV leaflets were thickened and incompetent; there was a multilobulated SOV aneurysm involving RCC burrowing into the IVS and a small outpouching of left coronary cusp. The surgery involved excision of the AV with the sinus, ligation of its penetrating portion at the crest of IVS, and closure of a 4 mm VSD at the upper end of IVS. It was followed by the replacement of the AV by a 23 mm bi-leaflet mechanical valve. He was discharged on Day 8 of surgery. Postoperative echocardiography (Figure 4A and B) showed the complete collapse of the IVS component of the SOV aneurysm, normally functioning mechanical AV and LVEF of 35%. He is doing fine at 5 months of follow-up, with mild improvement of LVEF to 40%.

## Discussion

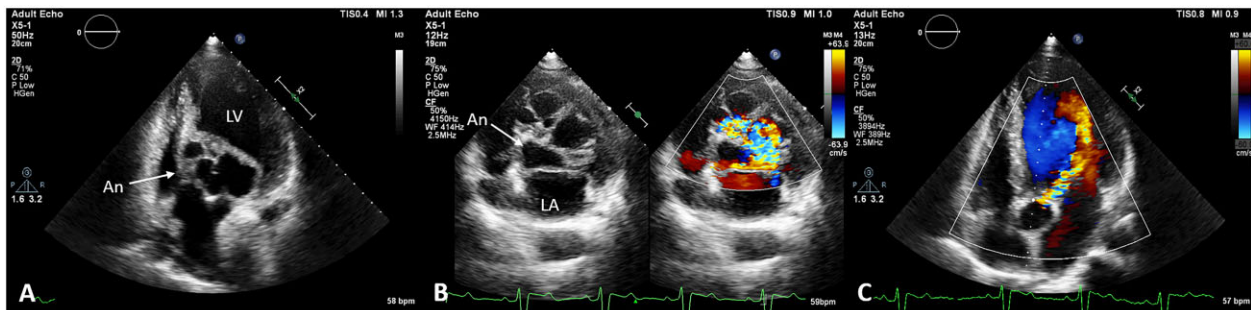
Sinus of Valsalva aneurysm (SVA) is a thin-walled outpouching of aortic sinuses. Right coronary cusp is most commonly involved (70–90%), followed by the non-coronary sinus (10–25%), and rarely left coronary sinus (<5%).<sup>1</sup> Congenital aneurysms are more common and are associated with various connective tissue diseases like Marfan's syndrome and Ehlers-Danlos syndrome, bicuspid AVs, VSDs, and aortic stenosis.<sup>1,9</sup> Acquired SVA is caused by atherosclerosis, endocarditis, Behçet's disease, ankylosing spondylitis, systemic lupus erythematosus, Takayasu's arteritis, chest trauma, or iatrogenic injury.<sup>9</sup>

Dissecting aneurysm into the IVS is a rare variant of SVA, first reported in 1947 by Warthen.<sup>10</sup> It is primarily congenital in origin and usually arises from RCC. The involvement of IVS is due to its proximity to the RCC, especially the middle part of it. Some authors believe that intraseptal extension is caused by intramural rupture of a congenital aneurysm and subsequent haematoma formation. It is actually a pseudoaneurysm as no true aneurysm wall was found during autopsy studies.<sup>2</sup> This pseudoaneurysm may gradually enlarge and rupture in one of the cardiac chambers.

In the literature, presentations of DAIS, described are as follows: congestive heart failure (53.8%), palpitations (30.8%), recurrent syncope (26.9%), chest pain (15.4%), sudden cardiac arrest (3.8%), infective endocarditis (3.8%), cerebral infarction (3.8%), and asymptomatic (3.8%).<sup>11</sup> Sinus of Valsalva aneurysm is associated with AR in 30–50% of cases. While mild AR is due to distortion of aortic cusp or aortic root due to long-standing volume overload, severe AR should be suspicious of aneurysm ruptured into the left ventricular outflow tract or endocarditis of AV.<sup>12</sup> It can also cause conduction disturbances varying from first-degree to complete heart block (CHB). It is due to



**Figure 1** Electrocardiogram showing normal sinus rhythm with bifascicular block.



**Figure 2** Two-dimensional echocardiography and colour Doppler images. (A) Dilated and loculated right coronary sinus and left coronary cusp in parasternal short-axis (PSAX) view. (B) Lobulated aneurysmal sinus of Valsalva burrowing into interventricular septum in apical four-chamber view. (C) Colour Doppler showing aortic regurgitation into left ventricle due to fenestration/rupture into left ventricle in apical 4-chamber (A4C) view.

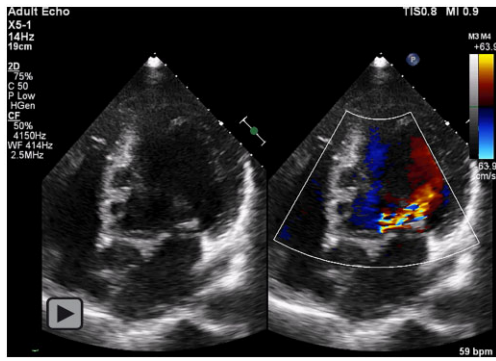
either the direct pressure effect of expanding the intraseptal aneurysm or low-grade localized inflammatory changes in the conduction tissue.<sup>11</sup>

Echocardiogram, CT, and cardiac magnetic resonance imaging are helpful in diagnosis. Colour Doppler shows continuous flow both in systole and diastole in ruptured SVA. Computed tomography angiography is the investigation of choice.<sup>13</sup> Cine cardiac magnetic resonance imaging, which can evaluate the

haemodynamics, identify the AR, quantify the shunt or turbulent, or fistulous blood flow, is considered the gold standard for diagnosis.<sup>14</sup>

Differential diagnoses of DIAS are hydatid cyst of the IVS, IVS abscess, or septal tricuspid valve aneurysm prolapsing into the left ventricle through a partially closed VSD.<sup>11</sup>

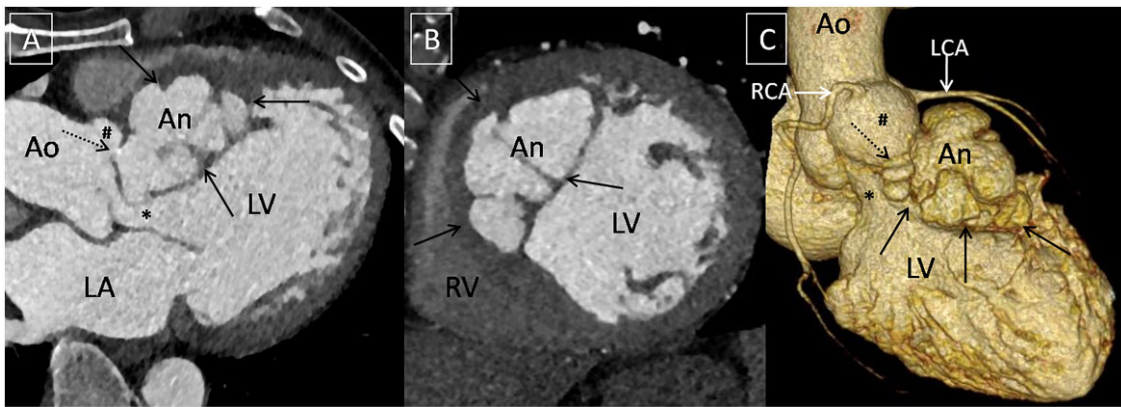
Surgical repair is the only option for DAIS. Medical management is only for temporarily stabilizing the patient until surgery. The first



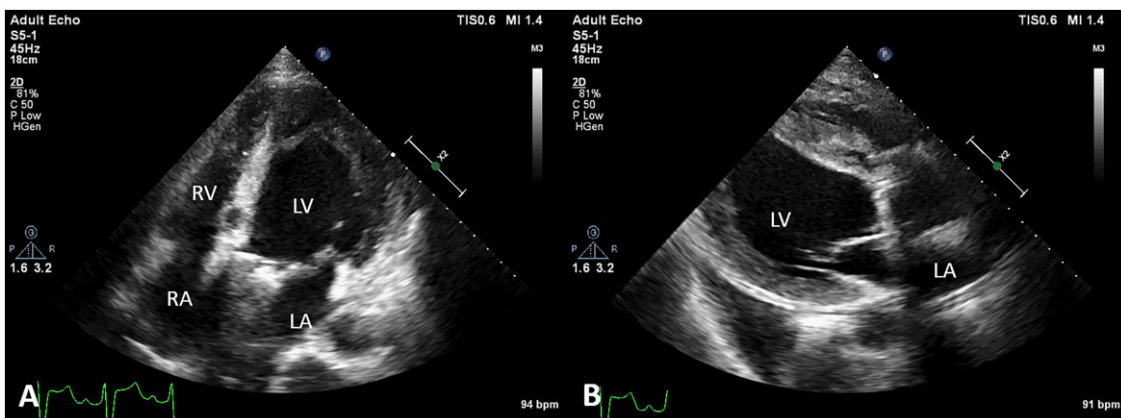
**Video 1** Apical five-chamber view showing lobulated aneurysmal sinus of Valsalva burrowing into interventricular septum rupturing into left ventricle and aortic regurgitation.

successful surgical repair of this condition was reported in 1976 by Heydem.<sup>15</sup> Dissecting aneurysm into the IVS has a progressive course and poor prognosis with a perioperative death rate of 3.9%. The perioperative complications include obstruction of the ventricular outflow tracts, arrhythmias, CHB requiring a permanent pacemaker, and communication with LV resulting in intractable heart failure.<sup>3</sup>

We believe that the index case had both congenital and acquired components contributing to the formation of DAIS. Our patient had a small subaortic VSD, which would have led to RCC prolapse and formation of SOV aneurysm. This would have led to both progressive AR and SOV aneurysms later on entering into the IVS. The multi-lobulated configuration of DAIS in the index case is very peculiar, and its rupture into LV is extremely unusual.



**Figure 3** Computed tomography angiography images (A–C) showing large multiseptated aneurysm (An) arising (dashed arrow depicting neck of the aneurysm) from the right sinus of Valsalva (#) and is seen burrowing into the basal and mid-cavity interventricular septum (black arrows) with mass effect on left ventricular outflow (\*). An, aneurysm; Ao, ascending aorta; LA, left atrium; LCA, left coronary artery; LV, left ventricle; RCA, right coronary artery.



**Figure 4** Postoperative images after excision of sinus of Valsalva aneurysm and replacement with a 23mm SJM mechanical valve in A5C view (A) and parasternal long-axis (PLAX) view (B).

## Lead author biography



Soumitra Ghosh is a third-year Senior Resident presently pursuing DM in clinical Cardiology at Post Graduate Institute of Medical Education and Research (PGIMER), Chandigarh, India.

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

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