

# Right Sinus of Valsalva Aneurysm and Dissection into the Interventricular Septum: A Case Report

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## ABSTRACT

Sinus of Valsalva aneurysm (SVA) is considered to be an uncommon cardiac anomaly, carrying a very high rate of mortality. After treatment, the prognosis is excellent. Thus, it is important to make a timely diagnosis and clarify the anatomical details of the SVA. Here, we report a right SVA with dissection into the interventricular septum, with bulging and incomplete rupture into the left outflow tract. The SVA was evaluated using echocardiography (transoesophageal, transthoracic, and three-dimensional echocardiography) and cardiac computed tomography (CT), especially three-dimensional reconstruction, to help us plan the surgical approach. After surgery, the patient's recovery was favorable.

## INTRODUCTION

Sinus of Valsalva aneurysm (SVA) is a rare cardiac anomaly that can be congenital or acquired. Both ruptured and unruptured SVAs can cause potentially fatal complications. The typical evolution of SVA is toward rupturing, and it carries a very high mortality rate. However, after treatment, the prognosis is excellent. Here, we report a right SVA and dissection into the interventricular septum with bulging and incomplete rupture into the left outflow tract. The SVA was evaluated using multiple diagnostic and imaging modalities. In particular, transthoracic echocardiography (TTE) and transesophageal echocardiography (TEE) were used simultaneously in one patient.

## CASE PRESENTATION

A 50-year-old male came to the emergency department with transient fainting. He was taking a walk and felt sudden, aggravated dizziness. He began to lose consciousness, called

out without receiving an answer, fell to the ground, and began to feel better a few minutes later. In the past month, he experienced dizziness without an obvious cause and syncope 3 times.

On arrival, he had no symptoms of heart failure. His blood pressure was 100/64 mmHg, with a heart rate of 75 beats/min and oxygen saturation of 99% in the emergency department. He had a surgical history of lumbar and leg fractures over 7 years prior. Electrocardiography showed a third-degree atrioventricular block. (Figure 1) The patient's hsTnT level was 0.040 ng/mg, which was higher than the normal range (0-0.014 ng/mg); CK-MB and MYO levels were within the normal range. His NT-ProBNP level was 1225.00 pg/ml, which was higher than the normal range (0-300 pg/ml), with left ventricular dysfunction. A head CT was negative, excluding a brain tumor. In the emergency department, the patient underwent pacemaker implantation, TTE and TEE at that time.

TTE revealed a right SVA and dissection into the interventricular septum. The SVA protruded into the left ventricular outflow tract in diastole and emptied in systole (Figure 2A-2E), with dilation of the left cavities and mild pericardial effusion. (Figure 2) The SVA was approximately 36 mm x 27 mm. The aneurysmal wall of the aortic sinus was close to the anterior mitral valve in the diastolic stage and far from the mitral valve in the systolic stage, with mild mitral regurgitation (vena contracta, 0.28 cm, area 2.7 cm<sup>2</sup>) and moderate aortic regurgitation (vena contracta, 0.26 cm, area 2.6 cm<sup>2</sup>). The left ventricular ejection fraction was 50% (Figure 2F). No ventricular or atrial septal defects were detected. TEE also revealed an aneurysm with the same features as that noted with TTE (Figure 3); however, unlike TTE, TEE clearly revealed the SVA-to-left ventricular outflow tract shunt, which was faintly visible on TTE (Figures 4A, 4B). (Figure 3) (Figure 4) The dilated aneurysm cavity filled the left ventricular outflow tract, resulting in stenosis, and a 5 mm wide defect was observed on the wall of the aneurysm near the anterior mitral valve, where blood flow was detected from the tum aneurysm cavity into the left ventricular outflow tract. Since the color flow jet was very close to the anterior mitral valve, the shunt flow was obstructed by the anterior mitral valve. A cavity of approximately 10x8 mm was seen in the interventricular septum, and blood flowed between the cavity and SVA through a fistula approximately 3 mm wide (Figures 4C, 4D). The right coronary valve prolapse and left coronary cusp were congenitally short. (Figure 5) The patient

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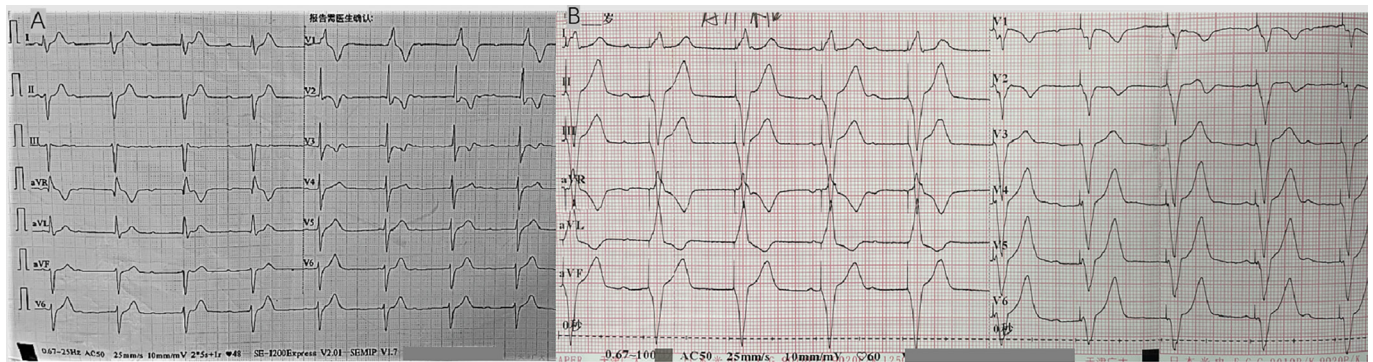


Figure 1. Initial evaluation. Twelve-lead electrocardiography with the evidence of third-degree atrioventricular block (A), after pacemaker implantation (B).

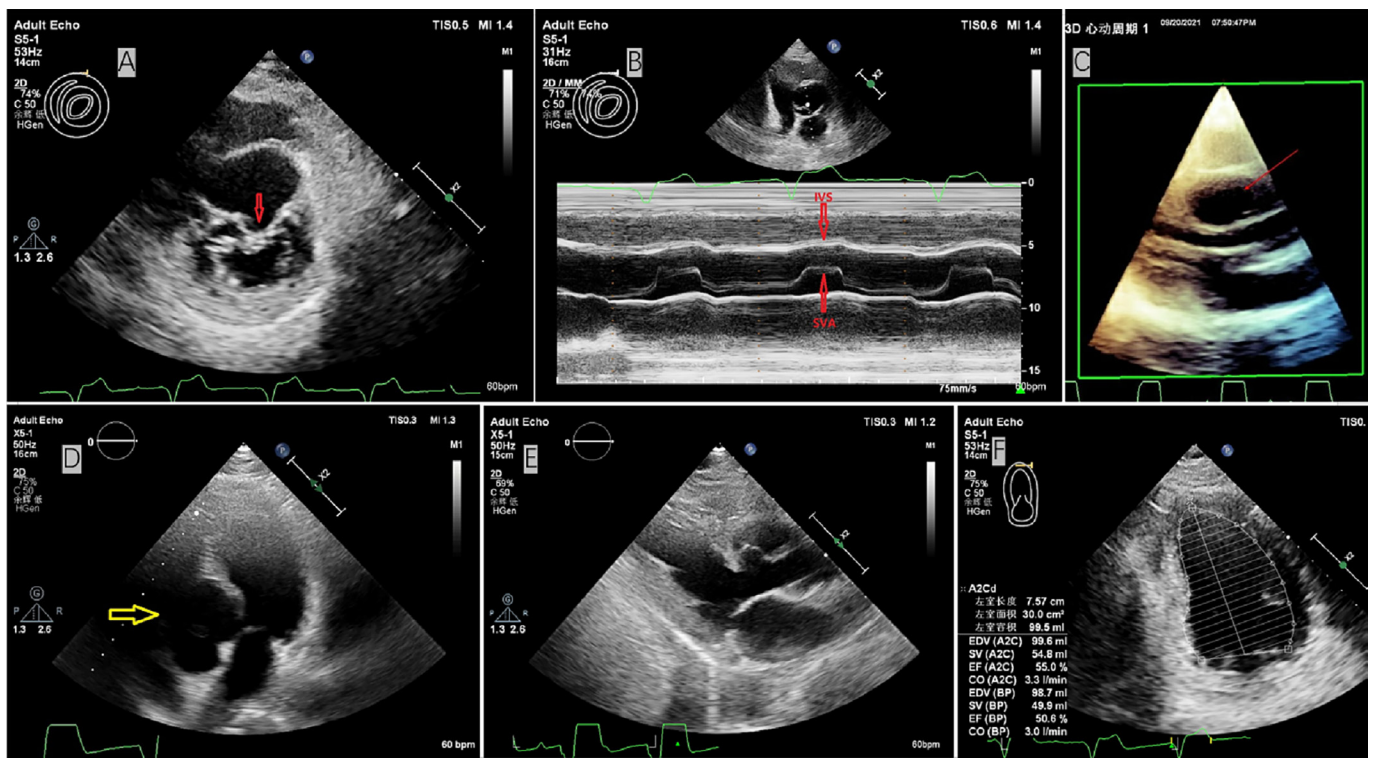


Figure 2. TTE showed right sinus of Valsalva aneurysm (red arrow) and dissection into the interventricular septum (yellow arrow). Short-axis parasternal view at the mitral valve level (A); M-mode imaging showing both systolic and diastolic aneurysm (B); three-dimensional TTE image (C); apical five-chamber view (D); parasternal long-axis view (E); left ventricular ejection fraction in two chamber views (calculated using the modified Simpson method) (F). TTE, transthoracic echocardiography; IVS, interventricular septum; SVA, sinus of Valsalva aneurysm

underwent a CT examination and received the same diagnosis. (Figure 6) An infectious disease workup was undertaken to rule out tuberculosis and syphilis. Genetic testing for connective tissue diseases, such as Marfan syndrome, was not undertaken because the patient did not exhibit symptoms of these diseases. The patient had no history of coronary intervention.

Therefore, surgical management was planned. After cardiopulmonary bypass was established and electromechanical arrest of the heart was achieved, the aortic valve was exposed using a transverse aortotomy. The intraoperative discoveries confirmed the above findings: a giant aneurysm

originating from the right aortic sinus protruding into the left ventricle outflow tract (Figure 7A). (Figure 7) The orifice of the protruding sacular aneurysm was closed with a running 4-0 Prolene suture and artificial mesh. The surgical view showed right aortic cusp prolapse and a short left aortic cusp (Figure 7B). Mechanical aortic valve replacement was performed. After weaning the patient from cardiopulmonary bypass, TEE was performed, revealing that the patch was in place and minimal mitral regurgitation was present. A closed "aneurysm" was left. For approximately 10 minutes, after protamine was given, TEE showed that the "anechoic" area

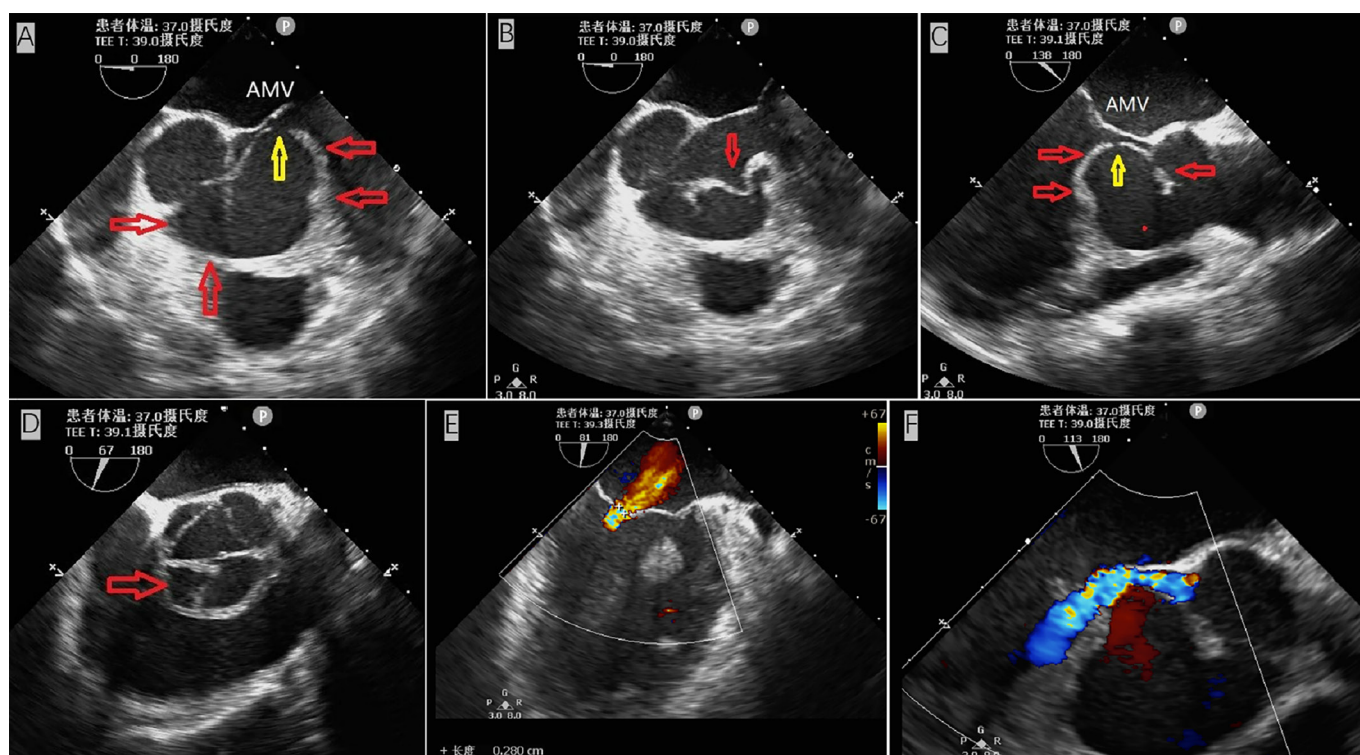


Figure 3. TEE showed right sinus of Valsalva aneurysm (red arrow) and incomplete rupture to the left outflow tract, resembling a fistula (yellow arrow). Non-standard 0 degrees four chamber view at midesophageal level in systole (A); nonstandard 0 degrees four chamber view at midesophageal level in systole (B); long-axis view at midesophageal level (C); aortic valve short axis view at midesophageal level (D); TEE with color Doppler revealed mild mitral regurgitation, vena contracta 0.28 cm (E); mild-to-moderate aortic regurgitation due to the prolapse of the right coronary cusp into the right sinus of Valsalva aneurysm (F). TEE, transesophageal echocardiogram; AMV, anterior mitral valve

in the “aneurysm” had turned into the weak echo, indicating that blood clotting had occurred in the “aneurysm.” (Figure 8) The patient had an uneventful postoperative course, and his recovery was favorable. The follow-up TTE performed 1 month after surgery showed a fully competent aortic valve with no regurgitation.

## DISCUSSION

SVAs are rare cardiac lesions that arise due to congenital or acquired aetiologies. Embryologically, SVA forms first as a blind diverticulum following pressure forces on the aortic root. The true prevalence of SVA is unknown. The estimated rate that has been cited in several studies dating back to 1839 is approximately 0.09% of the general population [Nguyen 2021; Weinreich 2015; Hope 1839]. Direct signs are an enlargement of the aortic root area between the aortic valve annulus and the sinotubular junction. Thus, the congenital defects that potentiate these pressure forces can lead to the development of an SVA, ranging from an asymptomatic presentation to severe aortic insufficiency and heart failure. The former may manifest as incidentally discovered unruptured aneurysms, and the latter may manifest as ruptured aneurysms [Hanna 2017]. In rare cases, unruptured SVAs may cause unusual structural and functional anomalies,

consequently presenting with atypical clinical manifestations, such as conduction disturbances [Agarwal 2018] and syncope [Matteucci 2009]. In this case, although a 3 mm wide defect was present, the aneurysm did not completely rupture. The right SVA protruded into the left ventricular outflow tract, and the patient developed symptoms of complete bundle branch block, syncope, and normal cardiac function. Most right coronary aneurysms occupy the right ventricle, even large (>5 cm) right coronary aneurysms [Pólos 2020].

The mechanism for heart block is explained by the extension of the SVA to the septal extension because the conduction tissue is located in the vicinity of the interventricular septum. The progressive evolution of aberrant conduction was described in a previous report of an aneurysm [Metras 1982; Hands 1985]. The small rupture and obstruction of the anterior mitral valve prevented the aneurysm from developing further and presenting symptoms, even though this case demonstrates the serious complications of a right sinus of Valsalva aneurysm dissecting the interventricular septum. Echocardiography, cardiac CT or CTA, and angiography are the most frequently used imaging modalities for the diagnosis of SVA.

TTE offered a noninvasive initial assessment of the morphology, location, and origin of the SVA. TEE was performed as a useful addition in cases of diagnostic uncertainty [Garrido 2002]. Ultrasound is of great value in the

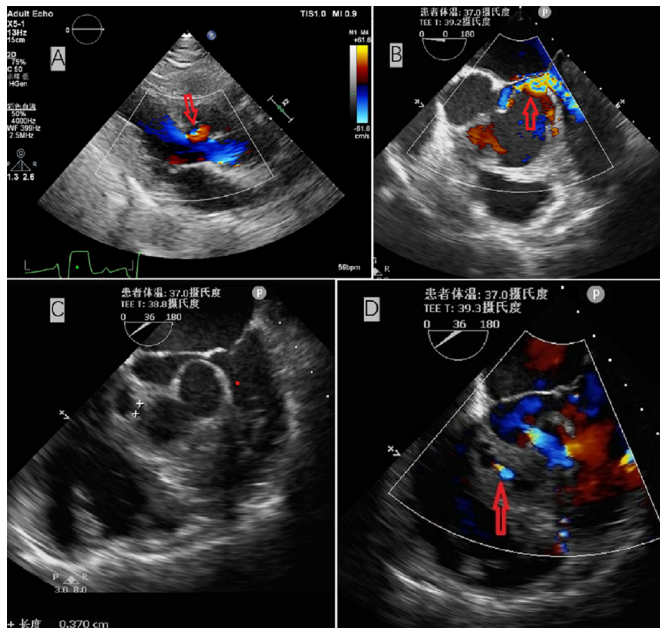


Figure 4. TTE and TEE with color Doppler, the arrow indicating the high-velocity jet from the right coronary sinus into the left ventricular outflow tract (A, B); a cavity was seen in the interventricular septum and blood flowed between the cavity and right sinus of Valsalva aneurysm through a fistula (+) of approximately 3mm wide. TTE, transthoracic echocardiography; TEE, transesophageal echocardiogram

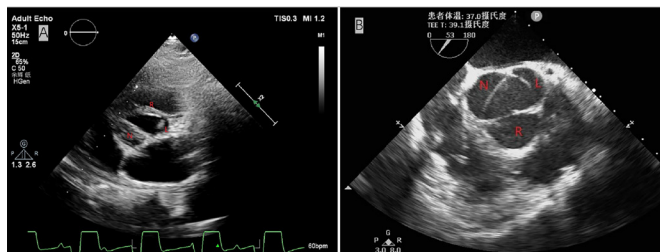


Figure 5. Left coronary cups was congenital short. TTE and TEE, aortic valve short-axis view at midesophageal level. TTE, transthoracic echocardiography; TEE, transesophageal echocardiogram

diagnosis of cardiac aneurysms, as previously reported by our team in the diagnosis of giant atrial septal artery aneurysm by ultrasound (TTE, TEE) [Mei 2021]. Three-dimensional echocardiography allowed for reconstruction of the SVA. In our patient, the TEE approach was more accurate than TTE in visualizing the defect and dissection into the interventricular septum. Regarding the detection of small defects, we faced a diagnostic dilemma with the transthoracic approach. With the interference of the anterior mitral valve, it was difficult to identify defects and color flow shunts using TTE. This case illustrates that TEE is frequently used to establish the diagnosis and provide more information concerning additional cardiac lesions in patients with an SVA. Aortic regurgitation should be evaluated after repair of the SVA by TEE.



Figure 6. Cardiac CT image showed the sinus of Valsalva aneurysm (red arrow) with extension into the interventricular septum (yellow arrow). Sagittal oblique view (A), preoperative three-dimensional reconstructed CT, with thin-section maximum intensity projection (MIP); right sinus of Valsalva aneurysm with extension into the interventricular septum at axial view (B); right sinus of Valsalva aneurysm with bulging into the left ventricular outflow tract (C). CT, computed tomography

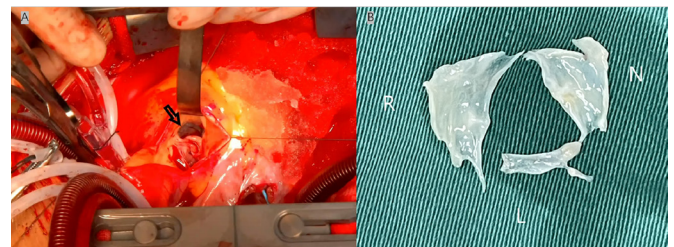


Figure 7. Intraoperative photography demonstrating the aneurysm (arrow) originating from the right aortic sinus. Sinus of Valsalva aneurysm (arrow) with an orifice in the right coronary annulus. Left coronary button is very close to aneurysm sac with a thin rim.

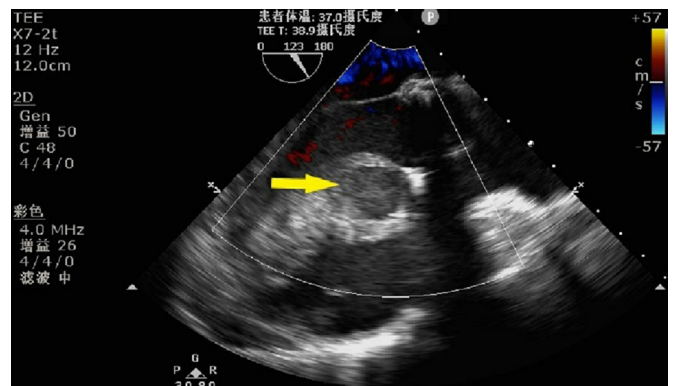
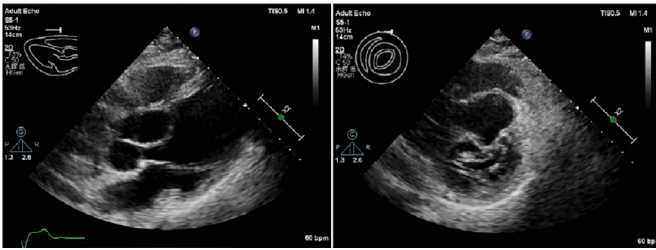
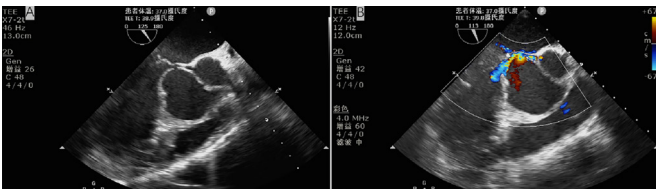


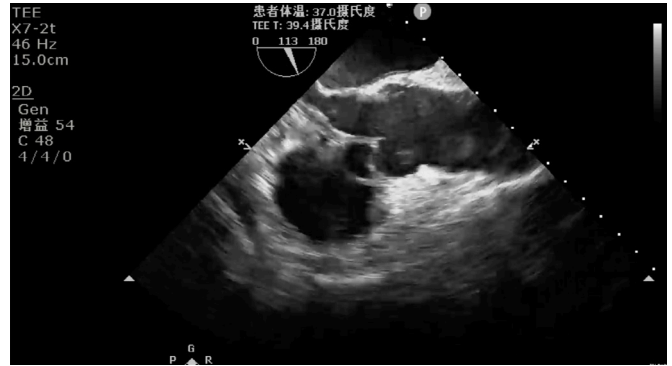
Figure 8. TEE with color Doppler. A closed "aneurysm" was left (arrow). The "aneurysm" had turned into weak echo, indicating that blood clotting had occurred.



Video 1. TTE showed the bulging and emptying of the right sinus of Valsalva aneurysm. Parasternal long-axis view (A) <https://vimeo.com/771200719>; parasternal short-axis view at the mitral valve level (B) <https://vimeo.com/771202371>. TTE, transthoracic echocardiography.



Video 2. TEE showed the right sinus of Valsalva aneurysm with a fistula to the left outflow tract, long-axis view at midesophageal level. Two-dimensional gray-scale view (A) <https://vimeo.com/771203735>; color flow Doppler (B) <https://vimeo.com/771204345>. TEE, transesophageal echocardiogram.



Video 3 TEE showed the right coronary sinus aneurysm with dissection into the interventricular septum, an echolucent cavity was seen in the interventricular septum. TEE, transthoracic echocardiography, long-axis view at midesophageal level. <https://vimeo.com/771210508>

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