



Laporan Kasus

RUPTUR ANEURISMA SINUS VALSAVA (RSOVA) PADA WANITA MUDA: LAPORAN KASUS

*RUPTURE SINUS OF VALSAVA ANEURISMA (RSOVA) IN YOUNG WOMAN: A CASE REPORT*Siti Tari Salsa,^{a*} Yusrina Saragih,^b^a General Practitioner, RSUD Muda Sedia, Aceh Tamiang, 24476, Indonesia^b Cardiologist, RSUD Muda Sedia, Aceh Tamiang, 24476, Indonesia

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A B S T R A K

Aneurisma sinus Valsava (SOVA) adalah kondisi jantung langka yang meliputi dilatasi abnormal akar aorta antara anulus katup aorta dan sambungan sinotubular yang disebabkan oleh kelemahan lamina elastis pada sambungan media aorta dan anulus fibrosus. Biasanya, sinus mencegah oklusi ostium arteri koroner pada saat sistol ketika katup aorta terbuka. Diameter sinus normal kurang dari 4 cm untuk pria dan 3,6 cm untuk wanita. Seorang perempuan berusia 28 tahun datang ke klinik rawat jalan dengan keluhan utama sesak nafas progresif saat aktivitas dan intoleransi olahraga selama satu bulan. Pemeriksaan fisik menemukan ascites, edema pitting bilateral, murmur kontinu di batas sternal kiri bawah, dan krepitasi di area basal paru. Foto toraks menunjukkan kardiomegali dengan peningkatan vaskularisasi, sedangkan elektrokardiografi menunjukkan sinus takikardia dengan deviasi aksis kanan. Ekokardiografi transtorasik (TTE) menunjukkan ruptur sinus valsalva yang berlawanan dari *Right Coronary Cuspid* (RCC katup aorta) menuju *RVOT* dengan diameter 16 mm dan gradien 72 mmHg, yang kemudian dikonfirmasi dengan ekokardiografi transesofageal (TEE).

A B S T R A C T

Sinus of Valsalva aneurysm (SOVA) is a rare cardiac condition that includes an abnormal dilatation of the aortic root between the aortic valve annulus and the sinotubular junction caused by weakness in the elastic lamina at the junction of the aortic media and the annulus fibrosus. Usually, the sinuses prevent the coronary artery ostia occlusion during systole when the aortic valve opens. The normal sinus diameter is less than 4 cm for men and 3.6 cm for women. A 28 years old woman admitted to ambulatory clinic with chief complaint progressive dyspnea on exertion and exercise intolerance in a month .Physical examination found with ascites, bilateral pitting edema, continuous murmur at the lower left sternal border, crackles in lower lung area . Chest x-ray showed cardiomegaly with increase vascularisation, whereas electrocardiography showed sinus tachycardia with right axis deviation. Transthoracic echocardiography (TTE) showed a ruptur sinus valsalva opposite from Right Coronary Cuspid (RCC of Aorta Valve) toward to RVOT with diameter 16 mm, gradient 72 mmHg, got confirmed by Transesophageal echocardiography (TEE)

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PENDAHULUAN

Sinus of Valsalva aneurysm (SOVA) is a rare cardiovascular anomaly marked by abnormal dilation of the aortic sinus, typically located just above the aortic valve. An uncommon consequence of sinus of Valsalva aneurysm is rupture. Ruptured sinus of Valsalva aneurysm is a rare cardiac anomaly and the incidence ranges from 0.23% to 1.5%.

Anatomically, a sinus of Valsalva is a pouch-like area in the aortic root, situated between the aortic valve ring and the sinotubular ridge. The left and right sinuses each house an opening for a coronary artery, while the rear sinus does not. These structures function to enable the aortic valve to open during heart contraction without blocking the coronary artery openings.¹

Ruptures of the sinus of Valsalva most commonly present between the ages of 20 and 40 and are rarely observed in pediatric or elderly populations. The condition shows a strong predilection for males (approximately 4:1 male-to-female ratio) and is more frequently reported in Asian populations than in Western groups.² Rupture occurs most often in the right coronary sinus (around 75% of cases), followed by the non-coronary sinus (about 20%), with the left coronary sinus being affected in only about 5% of cases.³

The location and direction of a sinus of Valsalva rupture largely dictate its clinical effects. Most commonly, a rupture from the right or non-coronary sinus into the right ventricle creates an abnormal connection between the aorta and either the right ventricular outflow tract or the right atrium. Ruptures from the left

coronary sinus are generally less significant, typically forming a passage to the left atrium or left ventricular outflow tract.³

The true incidence of this condition is not well-defined. Sinus of Valsalva aneurysms (SVA), which are the primary precursor to rupture, are estimated to account for roughly 0.1% to 3.5% of all congenital heart defects, equating to approximately 0.09% of the general population. However, only a minority of these aneurysms ever progress to rupture. An SVA may remain entirely asymptomatic until it ruptures, though in rare instances, it can cause cardiac arrhythmias, atrial fibrillation, or even complete heart block.⁴

Sinus of Valsalva Aneurysms (SVA) are most often present from birth, but can also develop later in life due to causes that damage the aortic wall, such as infections (syphilis, tuberculosis, endocarditis), atherosclerosis, or trauma. When congenital, SVA is commonly linked to other heart defects like a bicuspid aortic valve or aortic regurgitation.⁵

Symptoms of a ruptured sinus of Valsalva aneurysm can range from chest or abdominal pain to mild or severe shortness of breath. In many cases, it leads to acute heart failure, cardiac tamponade, or even sudden cardiac death. A holosystolic murmur is a typical finding during physical examination⁶.

However, imaging techniques are central to a correct diagnosis. Transesophageal echocardiography (TEE) is the primary method for detecting a ruptured sinus of Valsalva aneurysm (RSVA), though thoracic CT or MRI can also be useful to identify any additional abnormalities.⁵

CASE PRESENTATION

A 28 years old woman admitted to ambulatory clinic with chief complaint progressive dyspnea on exertion and exercise intolerance in a month .Physical examination found with ascites, bilateral pitting edema, continuous murmur at the lower left sternal border, crackles in lower lung area . Chest x-ray showed cardiomegaly with increase vascularisation, whereas electrocardiography showed sinus tachycardia with right axis deviation. Transthoracic echocardiography (TTE) showed a ruptur sinus valsalva opposite from Right Coronary Cuspid (RCC of Aorta Valve) toward to RVOT with diameter 16 mm, gradient 72 mmHg, got confirmed by Transesophageal echocardiography (TEE) .

A ruptured sinus of Valsalva aneurysm (SVA) is a rare cardiac abnormality, and associated with a severe left-to-right shunt if communicating with the right-sided heart chambers. Uncorrected, the rupture almost invariably causes deterioration in heart function. It was decided to refer the patient to get more advanced therapy with comprehensive facilities. Early surgical intervention is the choice of treatment to prevent further heart dysfunction. If the rupture occurs suddenly, hemodynamic decompensation may occur, leading to acute heart failure. The management of a ruptured SVA usually involves prompt cardiac surgical surgery. Surgical outcomes often show positive results, characterized by a favourable prognosis and modest recurrence rates.⁷



Figure 1. The sinus of Valsalva aneurysm ruptur with diameter 13-15 mm L-R Shunt,with VSD perimembranous subaortic with diameter 4-6 mm L-R Shunt on TEE



Figure 2. Chest x-ray showed cardiomegaly with increase vascularisation



Figure 3. electrocardiography showed sinus ryhtm with right axis deviation

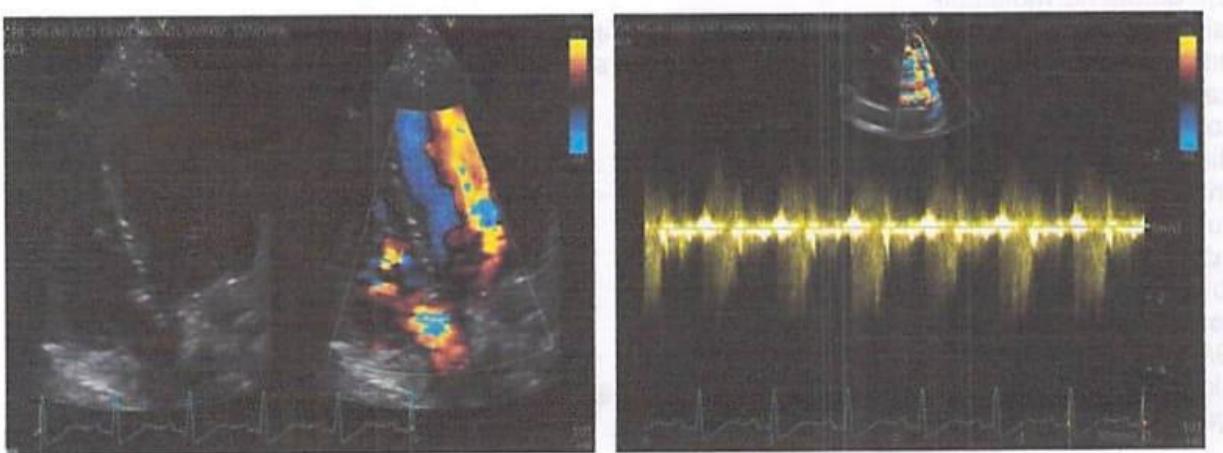


Figure 4. ecocardiography showed ruptur sinus valsalva opposite from Right Coronary Cuspid (RCC of Aorta Valve), systolic fubction good,EF 58%(BP Simon), diastolic function good, lvh konsentrik, RMWA(-)

DISCUSSION

A sinus of Valsalva aneurysm (SOVA) is a rare cardiac condition that includes an abnormal dilatation of the aortic root between the aortic valve annulus and the sinotubular junction caused by weakness in the elastic lamina at the junction of the aortic media and the annulus fibrosus. Usually, the sinuses prevent the coronary artery ostia occlusion during systole when the aortic valve opens. The normal sinus diameter is less than 4 cm for men and 3.6 cm for women.⁸ SoVAs can be congenital or acquired.⁹ Congenital SoVAs develop due to the absence of elastic lamina in the wall of the affected sinus, leading to an enlargement of the aortic root between the sinotubular ridge and the aortic valve annulus.^{10,11}

A SOVA most commonly occurs in the right coronary sinus (70%) and typically remains asymptomatic until it ruptures into nearby structures. The intracardiac aneurysmal sinus can rupture into an adjacent heart chamber, and the specific location of the rupture depends on the SOVA's position: the right, left, and

noncoronary sinuses are adjacent to the interventricular septum, left ventricular free wall, and interatrial septum, respectively. These anatomical proximities help explain the rupture's origin based on the SOVA's location.⁸

Ruptured sinus of Valsalva aneurysm is a rare cardiac anomaly and the incidence ranges from 0.23% to 1.5%.¹² This rupture creates a communication between the aorta and the heart chamber, leading to progressive heart failure. Without treatment, patients with a ruptured SOVA face poor prognoses and high mortality rates, making timely intervention essential. Any diagnosis of SOVA should prompt an urgent referral to a cardiothoracic surgeon for consideration of repair.¹³ SOVAs occur in approximately 0.2% to 0.9% of patients who have had cardiac surgery. SOVA can be either congenital or acquired; congenital SOVA, most commonly seen in men of Asian descent, results from abnormal development of the bulbus cordis and often coexists with anomalies like ventricular septal defect, aortic insufficiency, and bicuspid aortic valve.

Acquired SOVA can be linked to prior surgeries, atherosclerosis, endocarditis, syphilis, and other forms of trauma.⁸

The normal function of the sinuses of Valsalva is to prevent coronary ostia occlusion as the aortic valve opens during systole. Congenital and acquired etiologies for SOVA include connective tissue diseases (e.g., Marfan syndrome, Ehlers-Danlos syndrome), infections (e.g., syphilis, endocarditis, tuberculosis), trauma, chronic atherosclerosis, and cystic medial necrosis. The right coronary sinus is most commonly involved, followed by the noncoronary sinus and the left coronary sinus. Aneurysm enlargement can lead to disturbance of contiguous structures and result in complications such as arrhythmias or heart block, myocardial ischemia, or aortic regurgitation.⁴

Sinus of Valsalva aneurysm (SoVA) is an uncommon cardiac defect characterized by an abnormal dilatation of the aortic root between the sinotubular ridge and the aortic valve annulus. The most frequent complication of SVA is rupture, opening into the right atrium or ventricle or, as occurs in some acquired cases, the pleural or pericardial cavity. The diagnosis of ruptured SVA is relatively easy to establish because of the severe clinical picture that accompanies the condition. However, diagnosing a SVA before rupture is much less likely because it is usually silent. An unruptured SVA shows no clinical manifestations when it becomes infected, produces embolism, or compresses neighboring structures during its expansion. The most frequent complications are obstruction of the right ventricular outflow tract, aortic regurgitation, conduction disorders, and,

more rarely, myocardial ischemia due to compression of the coronary arteries.¹⁴

The frequency of Valsalva sinus rupture varies according to location, namely 60% in the right Valsalva sinus, 42% in the noncoronary sinus, and only 10% in the left Valsalva sinus. Rupture can occur spontaneously, after trauma, extreme physical exercise, or due to endocarditis.⁷ Rupture of a Valsalva sinus aneurysm primarily occurs into the right ventricle (60%), right atrium (29%), left atrium (6%), left ventricle (4%), or pericardium (1%).¹⁵ The pathophysiological consequences of this rupture depend on the volume of flow through the rupture, flow velocity, and the cardiac chambers involved. If the rupture occurs suddenly, hemodynamic decompensation may occur, leading to acute heart failure.⁷ Small, gradual, and progressive ruptures may be tolerated temporarily, occurring in 25% of cases.⁸ Clinical symptoms resulting from this condition include chest pain, shortness of breath, cough, and peripheral edema. If a rupture of the Valsalva sinus aneurysm is clinically suspected, diagnostic modalities such as TTE, TEE, MRI, and cardiac catheterization can be performed.¹⁶

Symptoms depend on the acuity and severity of rupture and can include chest pain, dyspnea, palpitations, or hemodynamic collapse. Diagnosis can be made with multimodality imaging. Traditionally, TTE and TEE have been the first-line imaging techniques, but cardiac CT (CCT) and cine CMR have gained recent popularity.¹⁷

For unruptured SoVAs, medical management is a temporary option for patients until definitive surgical intervention is possible. Medical therapy alone is insufficient as the optimal

treatment for unruptured SoVAs.¹⁸ Management includes a serial echocardiogram to assess aneurysm morphology and size. In addition, blood pressure control is essential. Medications such as beta-blockers, calcium channel blockers, angiotensin-converting enzyme inhibitors, and angiotensin receptor blockers should be used to lower blood pressure to the normal range and reduce aortic wall stress. Patients should also be advised to limit activities that may increase intrathoracic pressure and precipitate rupture, such as heavy lifting and strenuous exercises. In cases of acute rupture, medical stabilization with intravenous fluids and blood pressure support should be administered while managing heart failure or shock symptoms until surgical repair can be done.¹⁹

Surgical intervention is indicated for symptomatic, massive, or rapidly increasing unruptured SVA. Additionally, surgical management is recommended for unruptured SVAs that exhibit intraluminal thrombi, have a mass effect on adjacent tissues, or demonstrate recurrence. Surgical outcomes often show positive results, characterized by a favourable prognosis and modest recurrence rates. The management of a ruptured SVA usually involves prompt cardiac surgical surgery. However, surgical treatment can increase other risks and even cause death. Additionally, patients will often optionally for an interventional approach if it can achieve the same results as surgery. Nevertheless, percutaneous approaches have also been employed and have had favourable results.¹⁵ Based on a recent systematic review and meta-analysis, no statistically significant.²⁰

The diagnosis of SoVA requires a combination of history, physical examination, and imaging. The first line imaging study includes an echocardiogram to visualize the aortic root, identifying the aneurysm and any associated intracardiac abnormality. Additionally, cardiac studies such as CT and MRI can provide more precise information on the anatomic delineation of the aneurysm and its surrounding structures. Definite treatment of SoVAs includes surgery. However, TCC is an emerging technique used in managing both ruptured and unruptured SoVAs and is now increasingly preferred over traditional surgical approaches due to the reduction in surgical risk, shortened hospital course, and decreased recovery time.²¹

All surgical repairs are performed using cardiopulmonary bypass and cardioplegic arrest.²² A number of surgical techniques exist. The selection, however, is influenced by the aneurysm's dimensions, aortic valvular conditions like aortic insufficiency, the specific heart chamber affected, and any related intracardiac defects.^{23,24} The main surgical methods involve operating through the cardiac chamber into which the aneurysm has ruptured, through the aortic root via an aortotomy, or a dual approach combining an aortotomy with an incision into the affected heart chamber. Repair methods comprise primary and patch closure. Primary closure is frequently employed for small SoVAs, whereas patch closure is favored for larger ones. Employing primary closure for large SoVAs can deform the aortic sinus, leading to valve incompetence or high tissue tension at the repair site, potentially raising the risk of a future recurrent rupture. Surgical repair, in general, carries an

operative mortality rate as high as 3.6%, with survival rates nearing 90% at 15 years.²⁵

CONCLUSION

Ruptured aneurysm of right sinus of Valsalva is a rare cardiac anomaly. In most cases it usually ruptured toward right ventricle. The diagnosis of RSVA is challenging since it is rare. Anamnesa, physical examination, ECG and Echocardiography are very important for the diagnosis of sinus valsava. Early diagnosis and prompt surgical intervention can help reduce morbidity and mortality.

Prompt surgical repair is the recommended treatment to prevent the progression of cardiac dysfunction. A sudden rupture can lead to hemodynamic decompensation and acute heart failure. Therefore, the standard management of a ruptured sinus of Valsalva aneurysm (SVA) is immediate cardiac surgery. Postoperative outcomes are generally positive, with a good long-term prognosis and low recurrence rates.

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