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Case Report

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RUPTURED SINUS OF VALSALVA IN A NEONATE: CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT

Sinus of Valsalva aneurysm (SVA) is an abnormal dilatation of one or more aortic sinuses located between the aortic valve annulus and the sinotubular junction. Rupture of an SVA can lead to left-to-right shunting, most commonly into the right cardiac chambers. Although aortic sinus aneurysm rupture accounts for less than 1% of congenital cardiac lesions, its incidence varies by geography and population. Rupture of the sinus of Valsalva (RSOV) is a potentially fatal condition if not promptly diagnosed, and timely multidisciplinary management is essential for optimal outcomes. RSOV into the right cardiac chambers represents an uncommon complication characterized by atypical presentation, distinctive hemodynamic alterations, and unique echocardiographic features. We are herein presenting a rare case of a 13-day-old neonate striken with severe respiratory distress. Color Doppler echocardiography revealed a characteristic rupture of the sinus of right coronary cusp of the aorta, accompanied by a moderately sized atrial septal defect (ASD), and significant pulmonary hypertension.

KEYWORDS: Raptured sinus of valsalva aneurysm, Color doppler echocardiography for neonatal ruptured sinus of valsalva aneurysm, Neonatal sinus of valsalva aneurysm, Pulmonary hypertension.

INTRODUCTION

Sinus of valsalva aneurysm (SVA) is an uncommon congenital or acquired cardiac abnormality characterized by an enlargement of the aortic root segment between the aortic valve annulus and the sinotubular ridge, with an estimated prevalence of approximately 0.09% in the general population (Figures 1–3).[1]

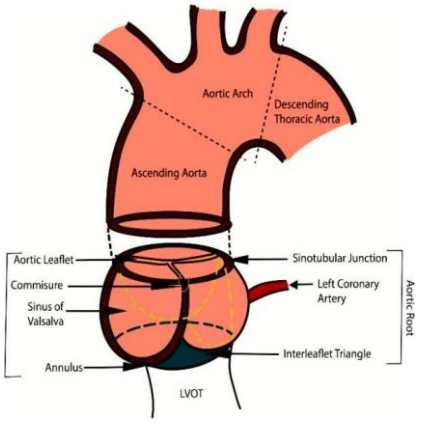


Figure 1: Illustration demonstrating the anatomy of the aortic root.

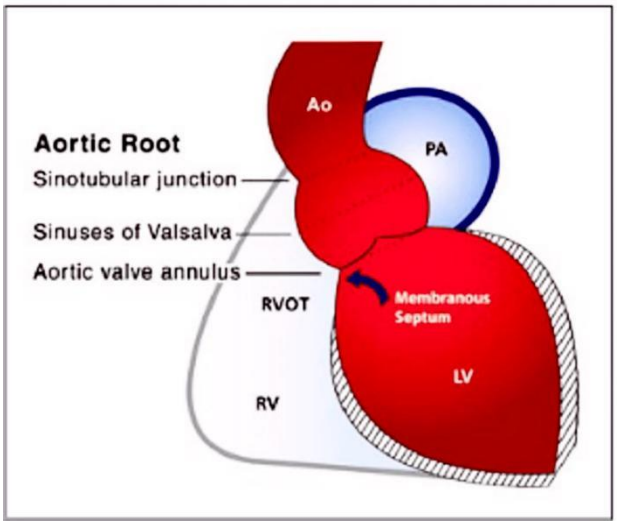


Figure 2: A diagrammatic representation of the aortic root in the coronal oblique plane illustrates normal anatomy and relationships. Ao: Aorta, PA: Pulmonary artery, RVOT: Right ventricular outflow tract, RV: Right ventricle, LV: Left ventricle.

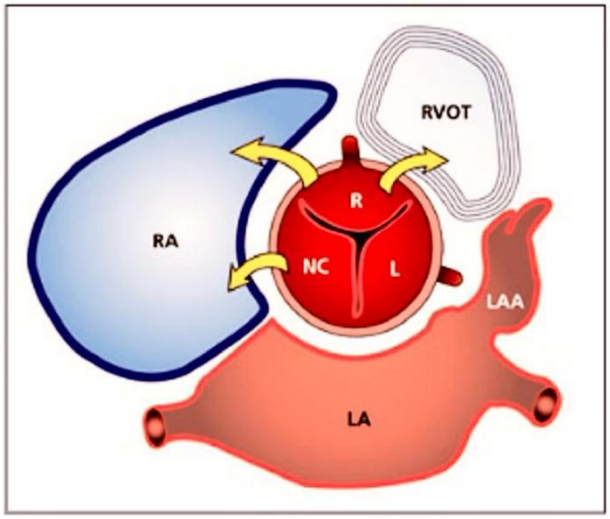


Figure 3: A diagrammatic representation of the sinus of Valsalva illustrates its relationship with adjacent cardiac chambers and the most frequent sites of rupture. RA: Right atrium, RVOT: Right ventricular outflow tract, LA: Left atrium, LAA: Left atrial appendage, R: Right coronary cusp, L: Left coronary cusp, and NC: Non-coronary cusp.

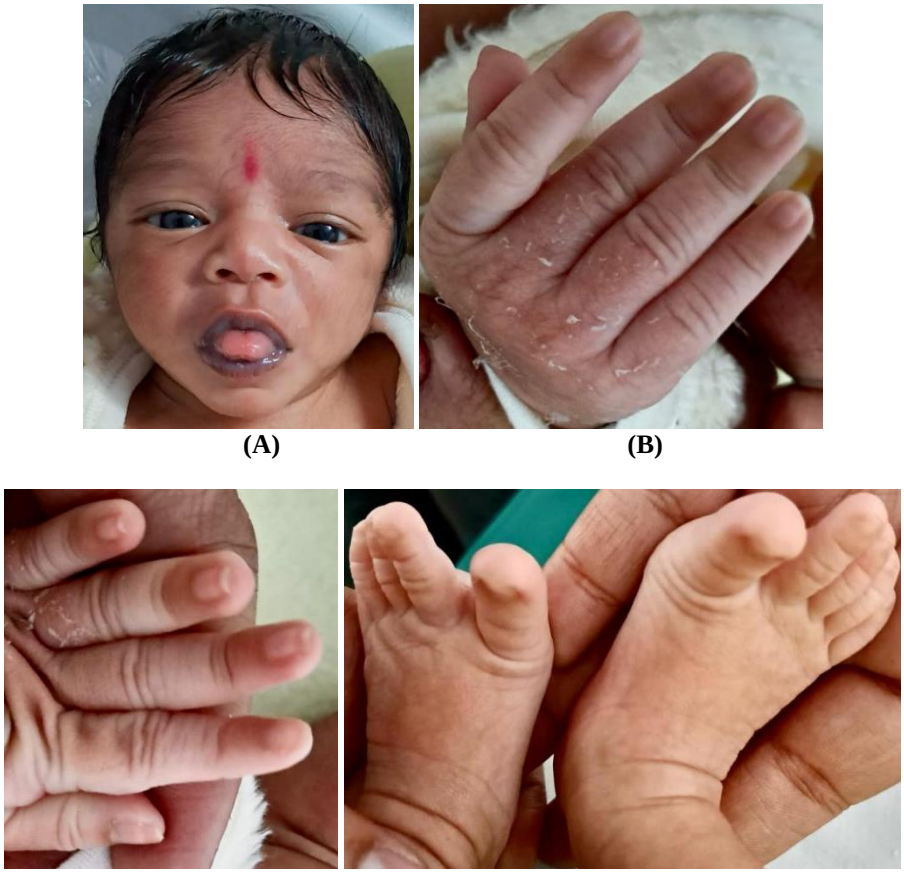
Ruptured aneurysms most frequently originate from the right coronary sinus (65–85%), less commonly from the non-coronary sinus (10–30%), and rarely from the left coronary sinus (<5%). The right ventricle (RV) serves as the most common receiving chamber, accounting for approximately 80–90% of cases.[1]

Several complications may arise, the most significant being rupture. The clinical signs and symptoms of RSOV vary according to the cardiac chamber into which the aneurysm ruptures—most commonly the right atrium (RA) or RV, and less frequently the left ventricle (LV). The presentation is typically acute, characterized by new-onset dyspnea and a continuous cardiac murmur in a previously asymptomatic patient. Diagnostic modalities include transthoracic echocardiography (TTE), transesophageal echocardiography (TEE), magnetic resonance imaging (MRI), cardiac catheterization, and cardiac computed tomography (CT). The condition is often misdiagnosed as a ventricular septal defect (VSD) due to the presence of a murmur and the anatomic location of the defect.

CASE REPORT

A 13-day-old male neonate was referred to us for a comprehensive evaluation of congenital heart disease. The parents reported a history of severe breathlessness since birth. The neonate was of average built but appeared markedly ill. There was notable puffiness of the face and eyelids, along with pronounced tachypnea. Distinct intercostal retractions were evident, accompanied by a prominent precordial bulge. No cyanosis or clubbing was observed (Figures 4A–D).

The neonate's weight was 2.7 kg, height 38 cm, blood pressure 70/60 mmHg, heart rate 141/min, respiratory rate 77/min, and SPO2 98% on room air. Cardiovascular examination revealed a grade 2/6 systolic murmur best heard in the left infraclavicular region. The second heart sound was normal. The remainder of the systemic examination was unremarkable.



(A) **(B)**

(C) **(D)**

Figure 4: Image of our index patient. (A) Puffiness of face and eyelids. (B) and (C) Right and left hand fingers were normal; (D) Both the toes were normal.

Chest X-ray (PA view) (Figure 5) demonstrated cardiomegaly with pulmonary plethora, suggestive of pulmonary edema.

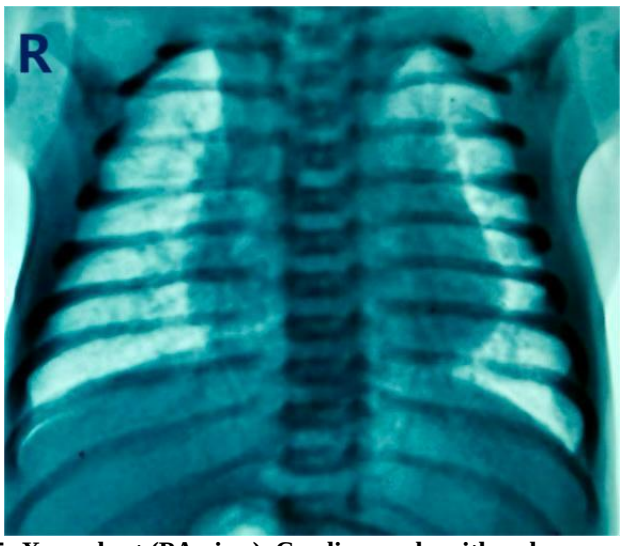


Figure 5: Xray chest (PA view). Cardiomegaly with pulmonary edema.

Resting ECG (Figure 6) revealed sinus tachycardia with features of left ventricular hypertrophy.

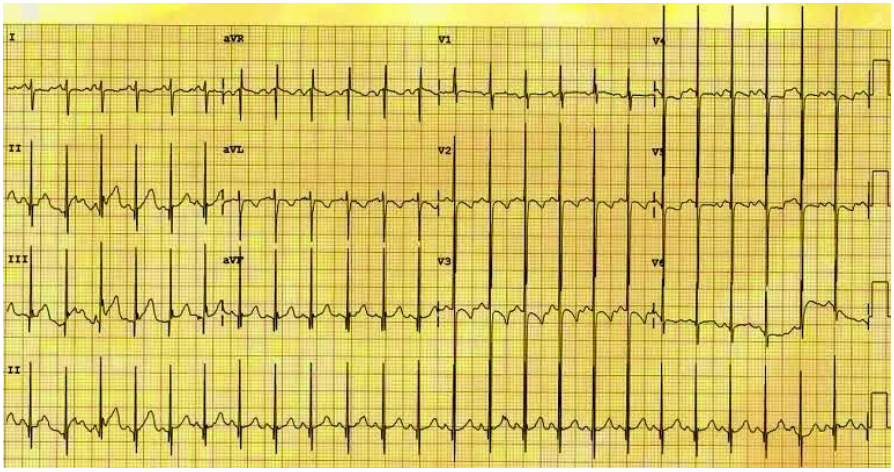


Figure 6: Resting ECG. ECG showed sinus tachycardia with left ventricular hypertrophy.

Transthoracic Echocardiography

The echocardiography system - My Lab X7 4D XStrain, Esaote, Italy, was utilized for all measurements and evaluations using a pediatric probe. Sequential segmental transthoracic echocardiography was performed in the classical subcostal, parasternal long-axis (PLAX), parasternal short-axis (PSAX), four-chamber (4CH), five-chamber (5CH), and suprasternal views.

M-mode Echocardiography

M-mode echocardiography of the left and right ventricles was performed, and the measured parameters are summarized in Table 1 and Figure 7.

Table 1: Calculations of M-mode echocardiography.

Measurements	**LV**
IVS d	3.4 mm
ID d	20.0 mm
PW d	2.4 mm
IVS s	6.1 mm
ID s	13.0 mm
PW s	6.3 mm
EF	67 %
% FS	35 %
EDV	12.8 ml
ESV	4.2 ml
SV	8.6 ml
Mass	8 g
IVS, interventricular septum, ID, internal dimension; PW, posterior wall, d, diastole; s, systole; FS, fractional shortening; EDV, end-diastolic volume; ESV, end systolic volume; SV, stroke volume; EF, ejection fraction.	

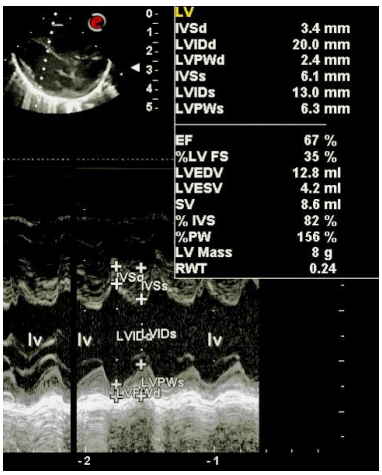


Figure 7: M-mode Echocardiography. LV volumetric estimation.

Summary of M-mode echocardiography

M-mode echocardiography demonstrated a borderline dilated LV with preserved systolic function. The LVEF was 67%, and the LV mass measured 8 g.

2-Dimensional-Transthoracic Echocardiography

Transthoracic echocardiography (TTE) was performed systematically using the sequential segmental approach (SSA). The echocardiographic findings observed are detailed below (Figures 8–16).

- Levocardia
- Situs solitus
- Concordant D-bulboventricular loop
- Atrioventricular (AV) concordance
- Ventriculoarterial (VA) concordance
- Normal relationship of the great arteries (NRGA)
- Left aortic arch with confluent pulmonary arteries
- Ruptured sinus of Valsalva (RSOV) of the right coronary cusp (large)
  - Size: 4.6 mm
  - Peak/mean gradient across RSOV: 10.1 / 3.9 mmHg
  - Left-to-right shunt
- Atrial septal defect (ASD) (moderate)
  - Size: 2.2 mm
  - Ostium secundum type
  - Peak/mean gradient across ASD: 15.2 / 4.7 mmHg
  - Left-to-right shunt
- QP/QS ratio = 2.2 : 1
- Mitral regurgitation: trace
- Borderline dilated LV with normal systolic function
  - LVEF: 67%
- Moderate to severe pulmonary hypertension
  - Estimated RVSP/PAP: 50 mmHg.
- No evidence of VSD, patent ductus arteriosus (PDA), coarctation of the aorta (COA), aortic stenosis (AS), or pulmonary stenosis (PS)

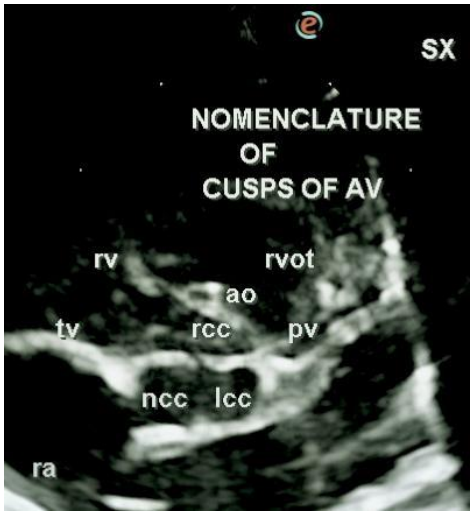


Figure 8: Nomenclature of the cusps of aortic valve. rv, right ventricle; ao, aorta; pv, pulmonary valve; rcc, right coronary cusp; lcc, left coronary cusp; ncc, non-coronary cusp; ra, right atrium; tv, tricuspid valve; rvot, right ventricular outflow tract; AV, arotic valve.

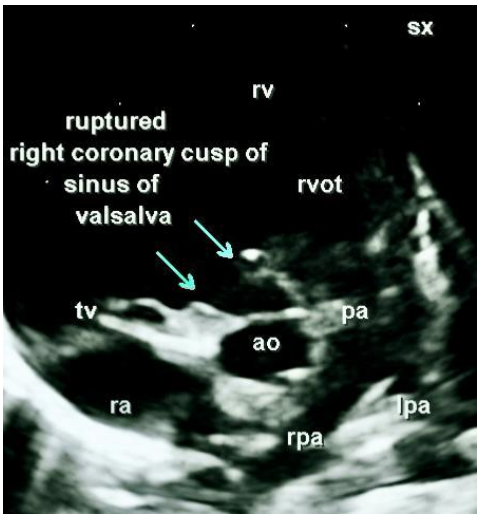


Figure 9: Rupture of sinus of valsalva of right coronary cusp. rv, right ventricle; ao, aorta; ra, right atrium; tv, tricuspid valve; rvot, right ventricular outflow tract; pa, pulmonary artery; rpa, right pulmonary artery; lpa, left pulmonary artery.

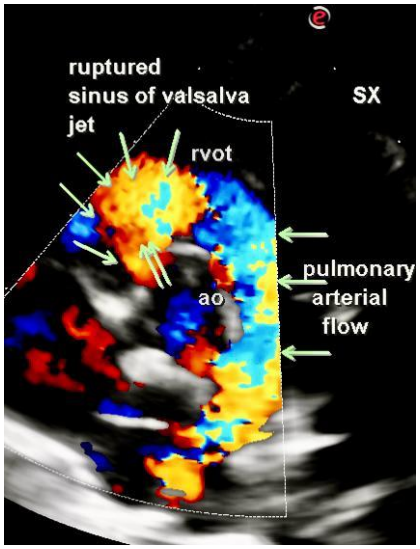


Figure 10: Color flow mapping of the jet of ruptured sinus of valsalva. rvot, right ventricular outflow tract; ao, aorta.

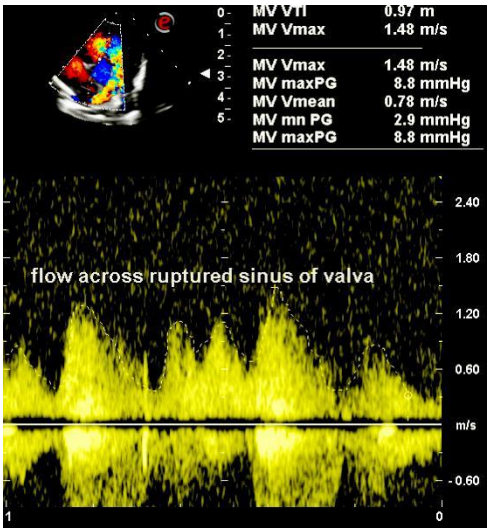


Figure 11: Continuous wave doppler imaging of flow across ruptured of sinus of valsalva. Peak/mean gradient across rupture sinus of valsalva was 8.8/2.9 mmHg.

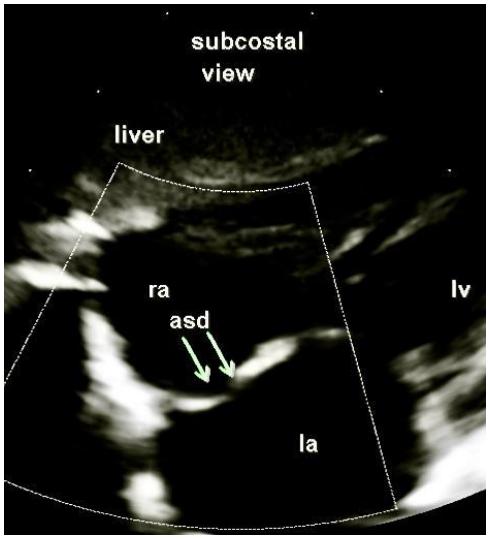


Figure 12: : Atrial septal defect. A moderate-sized (2.2 mm) ostium secundum ASD was identified. ra, right atrium; la, left atrium; lv, left ventricle; asd, atrial septum defect.

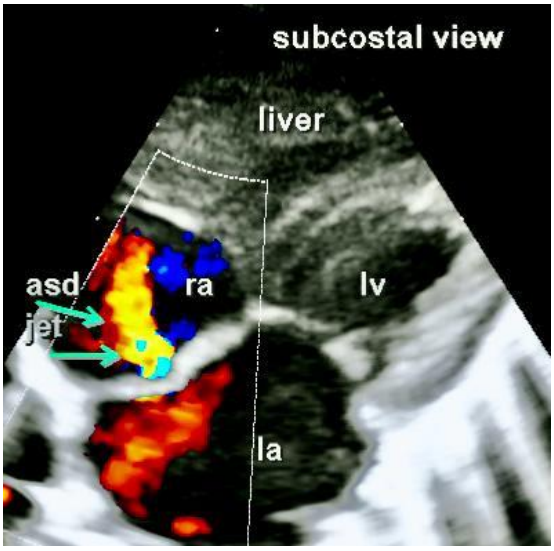


Figure 13: Color flow mapping across atrial septal defect. asd, atrial septal defect; ra, right atrium; la, left atrium; lv, left ventricle.

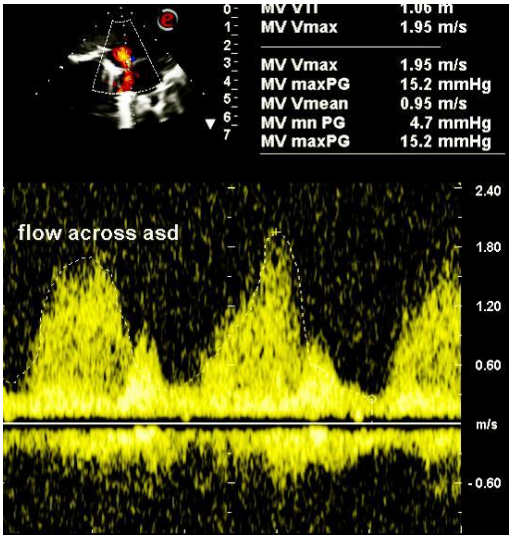


Figure 14: Continuous wave doppler analysis of flow across atrial septal defect. Peak/mean gradient across ASD was 15.2/4.7 mmHg.

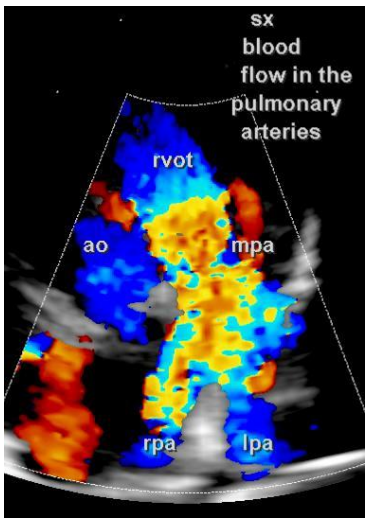


Figure 15: Color flow mapping of pulmonary arterial blow flow. Increased pulmonary blood flow was detected. rvot, right ventricular outflow tract; ao, aorta; rpa, right pulmonary artery; lpa, left pulmonary artery; mpa, main pulmonary artery.

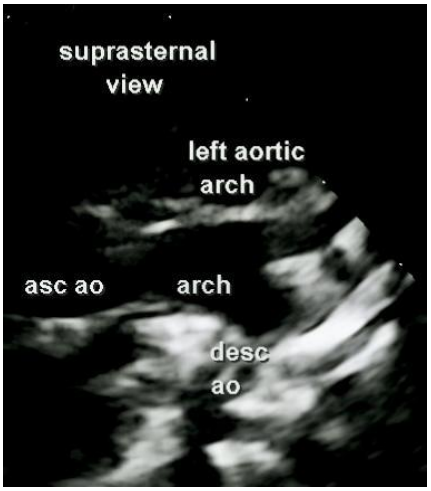


Figure 16: Left aortic arch. asc, ascending aorta; desc, descending aorta.

Summary of Transthoracic Color Doppler Echocardiography

Transthoracic color Doppler echocardiography confirmed a RSOV arising from the right coronary cusp of the aorta, with the rupture draining into the RV. A moderate-sized ostium secundum ASD was simultaneously identified, demonstrating a left-to-right shunt. Borderline LV dilation was noted, accompanied by moderately severe pulmonary hypertension.

DISCUSSION

Imaging Modalities for ROSV

TTE and TEE have traditionally been the primary diagnostic tools for SVA, but in recent decades, ECG-gated CTA and CMRI have gained increasing popularity for detailed anatomical assessment (Figures 17–20).[2, 3]

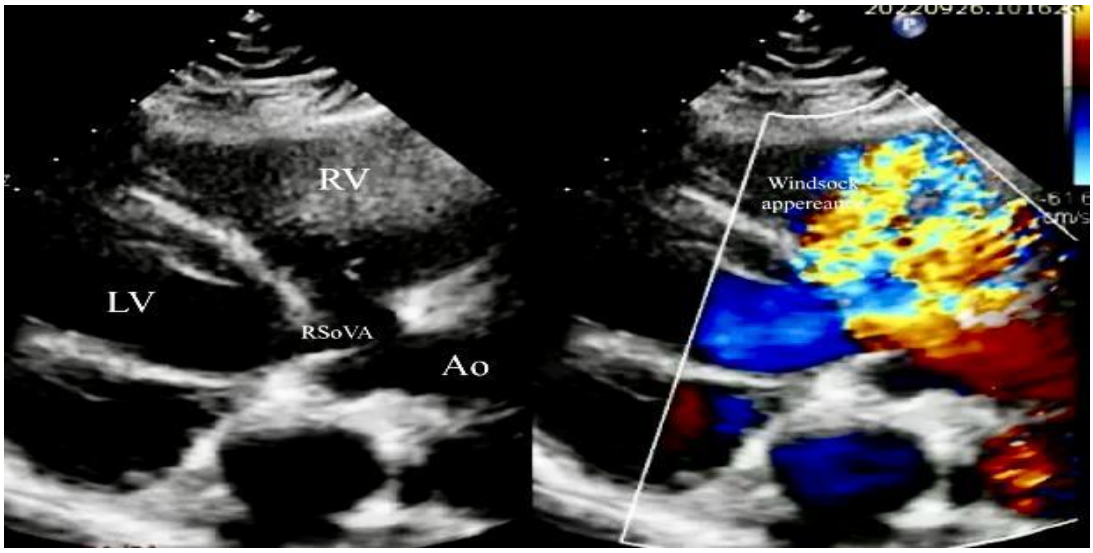


Figure 17: Transthoracic echocardiography identified a "windsock" appearance on parasternal long axis (PLAX) view. Ao = aorta; RV = right ventricle; LV = left ventricle; RSOVA: ruptured sinus of Valsalva aneurysm.

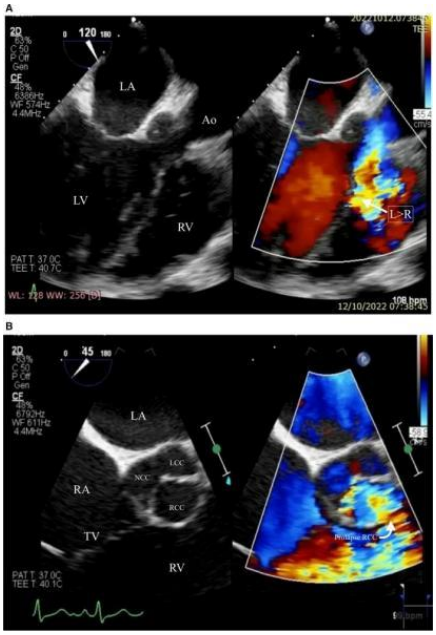


Figure 18: Transesophageal echocardiography confirmed rupture of the sinus of valsalva aneurysm on the Right coronary cusps (RCC). A and B Images taken flow from the RCC to the right ventricle on color doppler (arrow). Ao=aorta; RV = Right ventricle; LV = left ventricle; LA = left atrium; L = left; R = right; TV = tricuspid valve; NCC = non-coronary cusp; LCC=left coronary cusp; RCC=right coronary cusp.

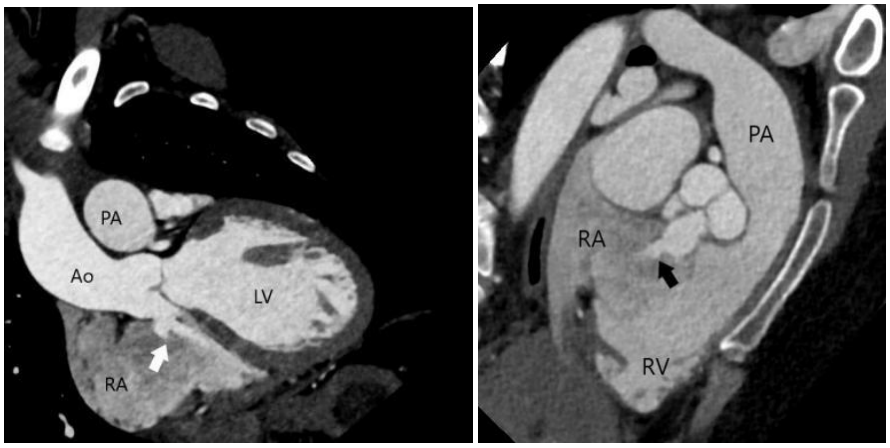


Figure 19: Orthogonal planes of gated cardiac CTA demonstrate a noncoronary sinus of Valsalva aneurysm that has ruptured into the right atrium (white arrow). A jet of contrast through the defect into the right atrium is also seen (black arrow).

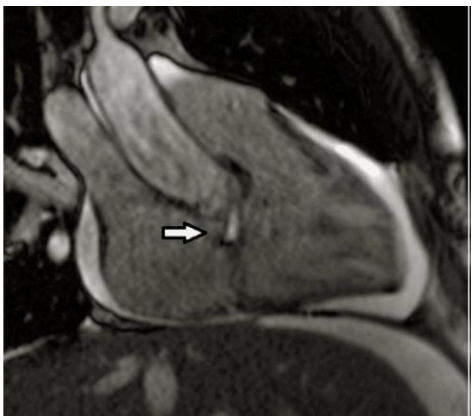


Figure 20: Cardiac Magnetic Resonance Imaging (MRI). Still frame of a modified right ventricular inflow-outflow steady-state free precession (SSFP) cine MRI, including the aortic root and showing a sinus of valsalva aneurysm rupture into the right atrium just above the plane of the tricuspid valve (arrow shows the jet entering the right atrium).

Multiplane transesophageal echocardiography (TEE) provides definitive diagnostic information regarding sinus of Valsalva aneurysm (SVA) and enables precise delineation of structural abnormalities as well as accurate localization of shunts for perioperative assessment.[4,5]

Two-dimensional TTE can identify up to 75% of patients with SVA [6,7]. Color Doppler imaging remains the modality of choice for detecting a ruptured aneurysm, while TEE, ECG-gated CTA, or cardiac magnetic resonance imaging (CMRI) are frequently required for diagnostic confirmation and comprehensive perioperative evaluation.

Typical TTE features of SVA include:

- Generalized enlargement of a single aortic sinus.
- A characteristic "wind-sock" extension arising from the body or apex of an otherwise normal aortic sinus upon rupture.
- Identification of associated defects such as ventricular septal defect (VSD), bicuspid aortic valve, and aortic insufficiency.

ECG-gated CTA and CMRI provide complementary, high-resolution three-dimensional anatomical data that are particularly valuable for pre-procedural planning and surgical guidance.

Background

SVA is an uncommon cardiac anomaly that may be congenital or acquired. John Thurnam first described this condition in 1840, and James Hope provided a more detailed description in 1939. The congenital form is usually clinically silent but may range from mild, asymptomatic dilation detected incidentally on two-dimensional echocardiography to symptomatic cases due to compression of adjacent structures or intracardiac shunting following rupture into the right-sided cardiac chambers. Approximately 65–85% of SVAs originate from the right sinus of Valsalva, whereas those arising from the non-coronary (10–30%) and left sinuses (<5%) are exceedingly rare.[8] SVA frequently coexists with aortic regurgitation and/or VSD.[2]

Pathophysiology

Congenital SVA results from dilation—usually of a single sinus—caused by a separation between the aortic media and the annulus fibrosus. A deficiency of normal elastic tissue and abnormal development of the bulbus cordis are implicated in its pathogenesis[4] (Figure 21).

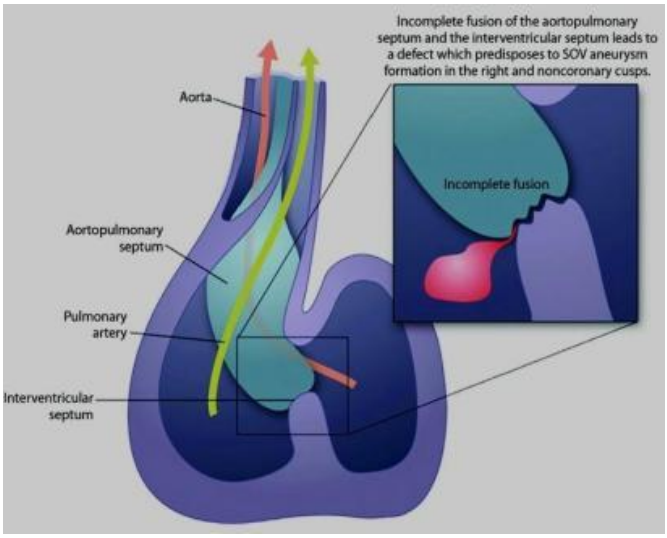


Figure 21: Illustration of the heart and root of aorta delineating the pathogenesis of congenital sinus of valsalva aneurysm.

Acquired forms arise from diseases affecting the aortic root such as atherosclerosis, syphilis, infective endocarditis, cystic medial necrosis, or chest trauma, which typically involve multiple sinuses. Rupture of the dilated sinus may establish an intracardiac shunt, commonly into the right atrium (Gerbode defect, ~10%) or directly into the right ventricle (60–90%). Cardiac tamponade may occur if rupture extends into the pericardial space.[9]

Etiology

Primary causes of SVA are congenital.

Secondary causes include:

- Atherosclerosis
- Syphilis
- Cystic medial necrosis or Marfan syndrome
- Blunt or penetrating chest trauma
- Infective endocarditis

Associated congenital defects include:

- VSD
- Aortic insufficiency
- Coarctation of the aorta

Epidemiology

United States data

SVA has been reported in 0.09% of cadavers in large autopsy series and in 0.14–0.23% of Western surgical

series.[10] Two-dimensional echocardiography has improved detection rates, and three-dimensional echocardiography offers additional diagnostic value.[11]

International data

The condition is more prevalent in Asian populations, with a reported incidence of 0.46–3.5%, correlating with a higher frequency of supracristal VSDs (~60%).[12]

Demographics

A male predominance exists, with a 4:1 male-to-female ratio encompassing both ruptured and unruptured forms. Unruptured SVA is often asymptomatic and incidentally detected, even in patients over 60 years of age. Most ruptured cases present between puberty and 30 years. A retrospective institutional review of 86 patients undergoing SVA repair (1956–2003) reported a median age of 45 years (range 5–80 years).[13]

Prognosis

The prognosis for SVA is poor in cases of progressive aneurysmal dilation or rupture unless timely surgical intervention is performed.[14] Actuarial survival for congenital SVA is approximately 95% at 20 years, as most do not rupture before that age. Unruptured aneurysms may remain stable for years but often progress or rupture. In a cohort of 86 surgically treated patients, ruptures occurred in 34%; six (7%) died perioperatively, and the 10-year survival rate was 63%.[13] Many required concomitant repair of associated VSDs, ASDs, or aortic valve lesions. Another series reported comparable outcomes, with 30-day postoperative survival of 92% among 65 patients aged 5–50 years, and 1-year survival in all remaining patients. Postoperative complications included septic shock due to endocarditis and paraprosthetic regurgitation.[15]

In a transcatheter series involving 25 patients, procedural success was achieved in 84%. One closure device embolized, two patients had residual leaks, and three required surgical intervention.[16]

Morbidity and Mortality

The natural history of SVA remains incompletely understood, as many cases present only upon development of complications.

Complications include:

- Expansion, rupture, heart failure, or sudden cardiac death
- Myocardial infarction secondary to coronary compression by an unruptured aneurysm
- Complete heart block due to compression of the conduction system
- Right ventricular outflow tract obstruction
- Infective endocarditis
- Cardiac tamponade following pericardial rupture
- Rarely, cerebrovascular embolic events

Congenital SVAs are frequently associated with additional structural abnormalities such as supracristal or perimembranous VSDs (30–60%), bicuspid aortic valve (15–20%), and aortic regurgitation (44–50%).[17,18]

Approximately 10% of patients with Marfan syndrome exhibit some form of SVA. Less common associations include pulmonary stenosis, coarctation, and ASD.

Rupture of SVA—leading to progressive heart failure, left-to-right shunting, or endocarditis—is the principal cause of death and rarely occurs before 20 years of age in congenital cases.

Medical Management

Medical management focuses on clinical stabilization, including optimization of therapy for heart failure and thorough perioperative assessment.[5] Transcatheter closure using Amplatzer devices has emerged as a minimally invasive alternative in selected patients, successfully avoiding sternotomy and cardiopulmonary bypass.[19–21]

Despite advances in percutaneous techniques, open-heart surgical repair, remains the gold standard of care. Surgical intervention also enables correction of coexisting lesions, such as aortic root replacement or ASD/VSD closure.[15,17,18]

CONCLUSION

The coexistence of a ruptured sinus of Valsalva aneurysm (RSOVA) with an atrial septal defect (ASD) is occasionally observed in patients presenting with severe dyspnea. Recognition of an associated ASD in individuals with RSOVA is crucial, as it substantially influences clinical management and therapeutic decision-making. This case report emphasizes the rarity of RSOVA coexisting with ASD in the general population. Echocardiography remains the cornerstone for accurate diagnosis, delineation of structural details, and assessment of hemodynamic impact. Timely diagnosis followed by early surgical repair is strongly recommended to prevent deterioration and improve clinical outcomes.

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