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Case Report

Successful Hybrid Management of Ruptured Sinus of Valsalva Aneurysm With Associated Coarctation of the Aorta and Bicuspid Aortic Valve in a Pediatric Patient

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ABSTRACT

Background: Congenital sinus of Valsalva aneurysm (SoVA) is a rare cardiac anomaly that predominantly affects the right coronary sinus. When ruptured, it can result in an aorto-cardiac shunt with hemodynamic consequences. Associated congenital lesions such as ventricular septal defect (VSD), coarctation of the aorta (CoA), bicuspid aortic valve, and patent ductus arteriosus may compound the clinical picture.

Case Presentation: A 12-year-old boy presented with acute chest pain, dyspnea, and palpitations. He was initially misdiagnosed with pneumonia and heart failure (New York Heart Association [NYHA] class III). Further evaluation revealed a congenital SoVA originating from the noncoronary cusp and rupturing into the right atrium (RA), producing a large left-to-right shunt. Transthoracic echocardiography demonstrated a windsock–like structure extending from the noncoronary sinus to the RA. Associated anomalies included severe discrete coarctation of the aorta.

Interventions: After stabilization with medical therapy, the patient underwent successful catheter-based CoA stenting. Because of the significant shunt and chamber dilation, open-heart surgery was subsequently performed.

Outcome: At postoperative follow-up, the patient reported no clinical symptoms. No residual coarctation or shunt was detected, and the boy remained hemodynamically stable.

Conclusions: This case underscores the importance of early recognition and comprehensive management of complex congenital cardiac anomalies, including SoVA, CoA, VSD, and valvular abnormalities. Multimodality imaging, catheter-based intervention, and surgical correction resulted in an excellent clinical outcome in this pediatric patient. ***(Iranian Heart Journal 2026; 27(1): 81-87)***

KEYWORDS: sinus of Valsalva; ventricular septal defect; coarctation of the aorta

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Congenital sinus of Valsalva aneurysm (SoVA) is an uncommon cardiac anomaly. It is more prevalent in Asian populations. It typically involves the right coronary sinus, followed by the noncoronary sinus and, rarely, the left coronary sinus. A nonruptured aneurysm is often asymptomatic, although it may gradually protrude and exert mass effect on adjacent structures. Ruptured sinus of Valsalva aneurysm results in the formation of a fistula between the aorta and a cardiac chamber, producing an aortocardiac shunt.[1-3] Rupture leads to a direct aortocardiac fistula, most often into the right atrium (RA) or ventricle, resulting in volume overload and congestive heart failure if untreated.[4] Isolated ruptures can be managed surgically; nonetheless, the presence of associated defects such as coarctation of the aorta (CoA), ventricular septal defect (VSD), or valve abnormalities poses diagnostic and therapeutic challenges.[5, 6]

We report a complex case requiring staged intervention to address life-threatening hemodynamic compromise.

Case Report

A 12-year-old boy was admitted to the emergency department with acute chest pain, dyspnea, dizziness, and palpitations after running up stairs. Initially, because of reduced functional capacity (NYHA class III), he was diagnosed with acute heart failure and pneumonia. Upon further questioning, his parents reported that since birth, he had experienced frequent respiratory infections, and their family physician had mentioned a probable cardiac defect. As he grew, respiratory infections occurred less frequently, and the parents believed he was improving; they did not seek medical management. Vital signs revealed tachycardia (130 beats/min), tachypnea (30 breaths/min), and bounding brachial and weak femoral pulses. Right arm blood pressure was 130/70 mm Hg and lower limb blood pressure was 90/60 mm Hg. On auscultation, reduced respiratory sounds over both lung bases, a grade 3/6 continuous murmur at the lower right sternal border, fixed splitting of the second heart sound, and a grade 3/6 systolic murmur at the left sternal border were detected. Transcutaneous oxygen saturation (SpO_2) was 94% on room air. On physical examination, short stature (height 134 cm, < 5th percentile) and low weight (27 kg, < 5th percentile) were noted. Laboratory tests were normal except for mild anemia (hemoglobin, 10.2 g/L). Chest radiography revealed an increased cardiothoracic ratio, right atrial and biventricular enlargement (Figure 1), increased pulmonary vascular markings, and blunted costophrenic angles. Electrocardiography (ECG) demonstrated RA enlargement, biventricular hypertrophy, and nonspecific ST-T changes. Transthoracic echocardiography revealed dilation of all cardiac chambers, particularly the RA, with preserved left ventricular systolic function. A thin-walled saccular lesion arising from the noncoronary sinus of the aorta, extending into the RA near the tricuspid annulus and forming a dark space with a windsock appearance, was noted. The sac fused with and displaced the tricuspid valve. Color Doppler imaging showed turbulent flow within the aneurysm and at the perforation site. Severe tricuspid regurgitation (TR), moderate aortic insufficiency, and narrowing of the aortic isthmus were also noted. A faint high-pressure gradient from the left ventricle to the right ventricle suggested a previous VSD, which was nearly closed because of prolapse of the aortic cusp (Figures 2-4).

After a few days of medical treatment and patient stabilization, catheterization and coarctation balloon angioplasty were performed (stage 1 of treatment). Cardiac catheterization confirmed severe discrete CoA with multiple collateral vessels;

ruptured sinus of Valsalva (SoVA) to the RA; severe TR, aortic runoff, and mild pulmonary hypertension; and normal arterial oxygen saturation (SpO_2 = 96%). CoA stenting was performed using an 8 × 30–mm Cook Formula stent (Figures 5 and 6). Immediate gradient relief and improved femoral pulses were noted after balloon CoA angioplasty (Table 1).

Table 1. Cardiac catheterization pressures before and after CoA stenting

Chamber	Prestenting (mm Hg)	Poststenting (mm Hg)
AAO	125/85 (105)	115/75
Femoral Artery	110/75 (88)	N/A
DAO	80/55	120/80
RA, RV, PA	Not documented	Normalized

CoA: coarctation of the aorta; AAO: ascending aorta; mm Hg: millimeters of mercury; DAO: descending aorta; RA: right atrium; RV: right ventricle; PA: pulmonary artery; N/A: not applicable

Two weeks poststenting, definitive surgical correction was proposed because of severe RA dilation and a substantial left-to-right shunt. The patient underwent open-heart surgery (stage 2) comprising repair of the ruptured SoVA to the RA, tricuspid valve repair, aortic valve repair, atrial septal defect surgical closure with a pericardial patch, and patent ductus arteriosus ligation. Postoperatively, echocardiography revealed no residual shunt, mild aortic insufficiency, and mild TR. The patient was discharged in stable condition (NYHA functional class I) and advised to undergo follow-up echocardiography. At routine outpatient follow-up, he reported no further symptoms, and resolution of cardiac murmurs was noted on physical examination.

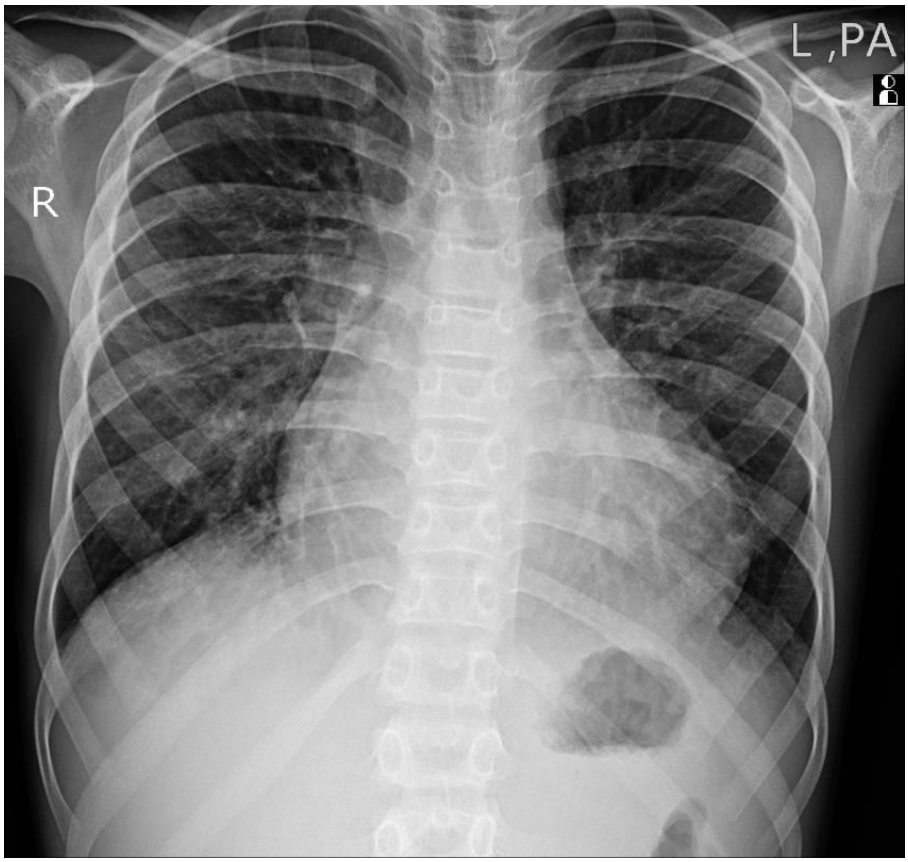

Figure 1. Chest radiograph showing increased pulmonary vascular markings and cardiothoracic ratio

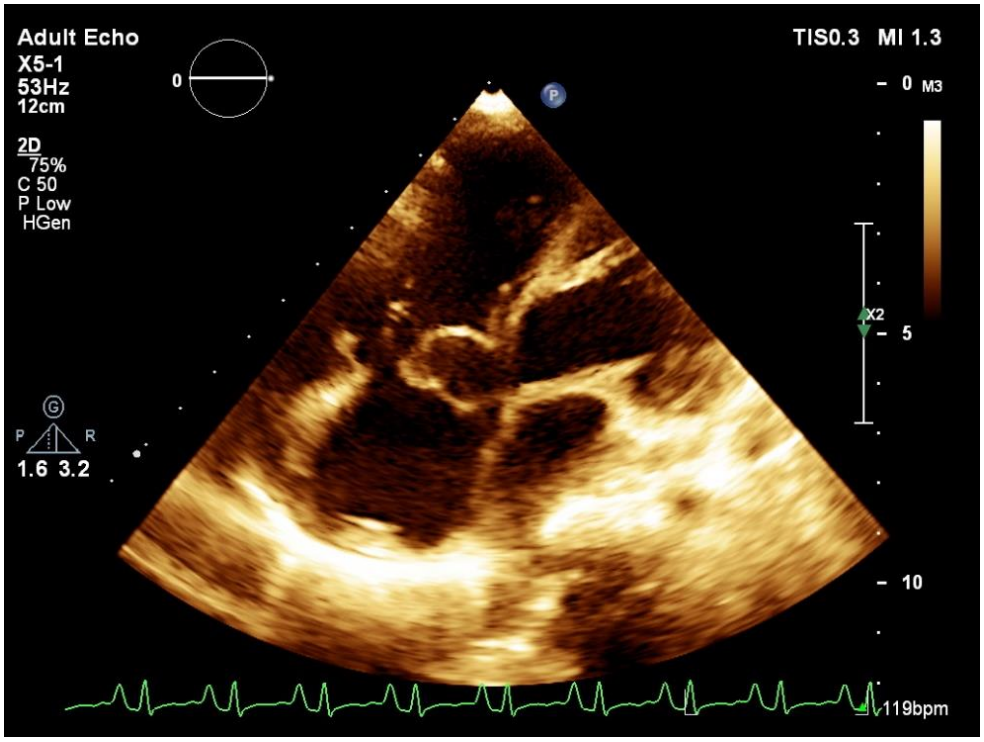

Figure 2. Two-dimensional echocardiography

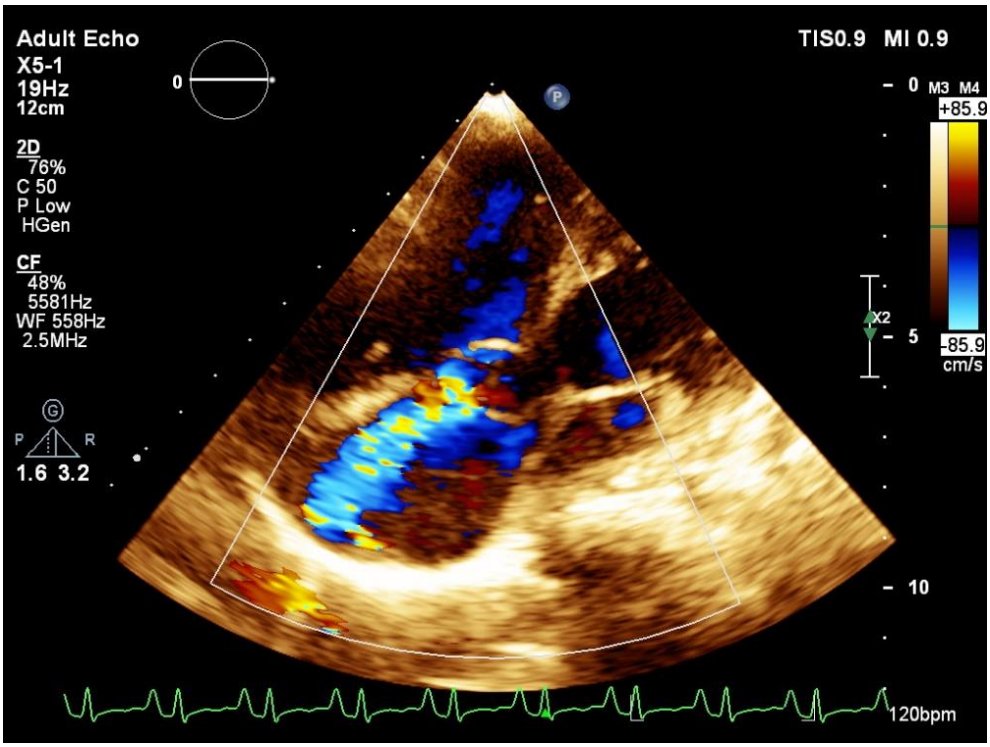

Figure 3. Color echocardiography demonstrating rupture of the sinus of Valsalva into the right atrium

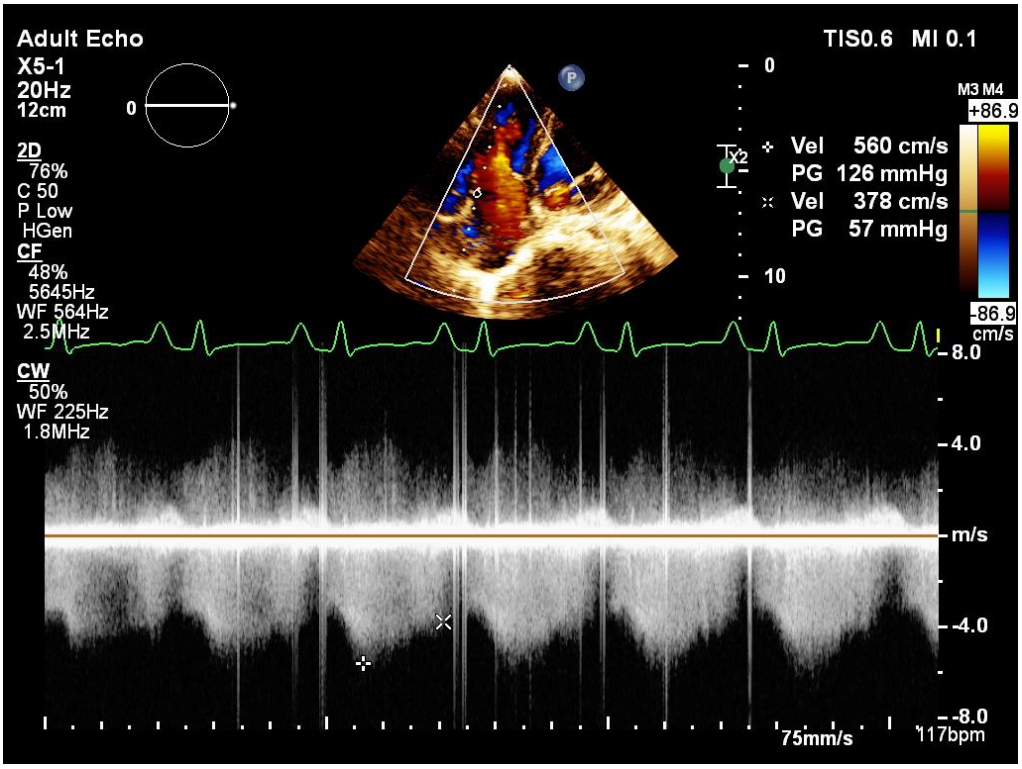

Figure 4. Doppler echocardiography showing a high-pressure gradient across the defect

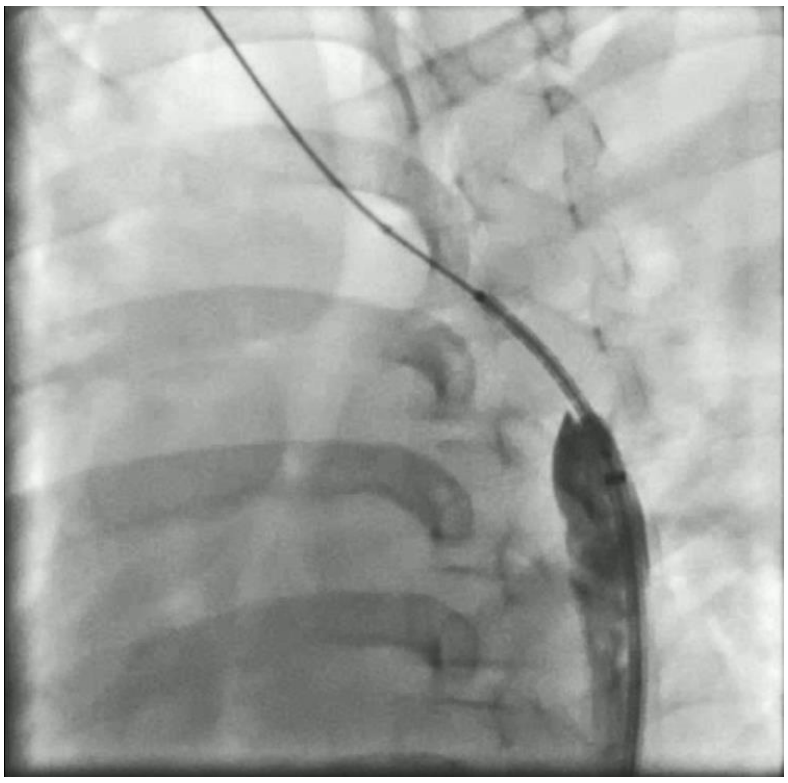

Figure 5. Coarctation of the aorta before stenting

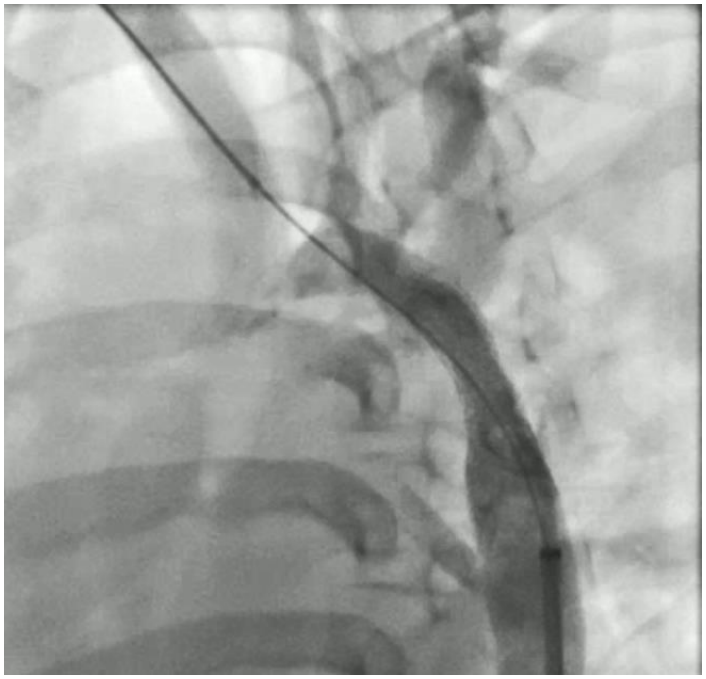

Figure 6. Coarctation of the aorta after stenting (one collateral vessel visualized)

DISCUSSION

Congenital SoVA, particularly into the RA, is rare and often associated with other cardiac defects such as bicuspid aortic valve, CoA, and residual VSD.[7-9] The patient's clinical presentation-continuous murmurs, differential pulses, and signs of right heart overload-was classic; nevertheless, delayed diagnosis due to physiologic adaptation masked the severity of the disease for years.

Echocardiography remains the cornerstone of diagnosis, especially with Doppler visualization of abnormal flow. Multimodality imaging and catheterization allow precise anatomic assessment for stenting and surgery.[10-12]

Staged interventions in this case, initial CoA stenting followed by surgical correction, enabled hemodynamic stabilization and optimized outcomes. Residual defects were minimal postoperatively, and the patient maintained good ventricular function.

This case illustrates the importance of multimodality imaging and staged management in congenital heart disease. The patient's long-standing symptoms and misinterpreted early signs emphasize the need for early screening in suspected congenital defects.

CONCLUSIONS

This case exemplifies the complex interplay of multiple congenital cardiac anomalies presenting with SoVA rupture. Timely diagnosis, a stepwise interventional strategy, and comprehensive surgical repair resulted in a favorable clinical recovery. Pediatric patients with congenital SoVA rupture warrant careful evaluation for associated defects and should be managed in specialized centers with expertise in congenital cardiac surgery.

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