

CASE PRESENTATION

A rare acute right heart failure: ruptured sinus of Valsalva into the right atrium

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Abstract: Sinus of Valsalva aneurysms (SVA) are rare, mostly congenital, cardiac abnormalities. SVA rupture creates an aortico-cardiac fistula, being most common on the right side: right coronary sinus ruptured into right atrium/ventricle. We report the case of a 28-years-old man which was referred to hospital due to right upper quadrant pain without irradiation and emphasizing a dynamic dyspnea and fatigue. The clinical examination outlines a right heart failure and the echocardiographic assessment emphasized the rupture of sinus of Valsalva aneurysm (RSVA) into the right atrium. The anomaly was corrected surgically with excellent outcome.

Keywords: sinus of Valsalva, rupture, aneurysm, right atrium, echocardiography

Abstract: Aneurismele de sinus Valsalva (SVA) sunt anomalii cardiace rare, cel mai frecvent congenitale. Ruptura de SVA creează fistulă aorto-cardiacă, fiind mai frecvent întâlnită la nivelul cavităților cordului drept: sinusul coronar drept rupt în atriu/ventricul drept. Prezentăm cazul unui bărbat în vârstă de 28 de ani care s-a internat pentru durere toracică fără iradiere, asociind dispnee și fatigabilitate. Examenul clinic evidențiază insuficiență cardiacă dreaptă, iar evaluarea ecocardiografică a evidențiat ruptura aneurismului de sinus Valsalva (RSVA) în atriu drept. Anomalia a fost corectată chirurgical cu un rezultat excelent.

Cuvinte cheie: sinus Valsalva, ruptura, aneurism, atriu drept, ecocardiografie

INTRODUCTION

SVA can be classified as congenital and acquired: the defects are due to lack of media on the transition area (*weakness area*) between the fibrous ring of the aortic valve and aortic isthmus. Sometimes aneurysms can coexist with other congenital cardiac malformations such as: a ruptured right sinus of Valsalva into the right ventricle through a ventricular septal defect¹, permeable ductus arteriosus, atrial septal defect, subaortic stenosis and tetralogy of Fallot²⁻⁴. SVA had an incidence ranging from 0.1 to 3.5% of all congenital heart defects⁵, 0.14–0.96% of open heart surgery cases⁶ and a prevalence of 0.09% in the general population on an autopsy series⁷.

Acquired aneurysms can develop as a result of atherosclerosis, endocarditis, aortic dissection, thoracic trauma, syphilis, Behçet's disease, Marfan syndrome, Loey-Dietz syndrome, Ehlers-Danlos syndrome, cystic media necrosis^{3,4,6,8}.

Most frequently SVA involve the right coronary sinus (RCS, 65–85%), non-coronarian sinus (10–30%)^{9,10} and left coronary sinus (below 3%)¹¹. Extracardiac RSVA are rare. Some authors noticed the difference between Asian and Western patients with RASV: in Asian was found a higher RASV incidence, more aneurysms originating from the right coronary sinus, more aneurysms rupture into right ventricle, a higher incidence of association with ventricular septal defect and lower incidence of association with bicuspid aortic valve¹².

SVA can progress silently for several years and rupture occurs in 3-4 decade of life usually into an adjacent chamber (right heart cavities)¹³, most commonly into right ventricle (86.7%)¹⁴, or into the right atrium, even bicameral rupture into right atrium and right ventricle¹⁵, into interventricular septum generating third degree atrioventricular block or into pericard producing tamponade^{11,16-18}. Initially rupture may be silent, but later manifests itself as a progressive heart failure

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caused by arterio-venous communication and aortic regurgitation¹⁹.

Sinus of Valsalva aneurysm classification was proposed by Sakakibara and Konno (1962)²⁰:

- Type I: aneurysm has its origin in the left side of the right coronary sinus (RCS) and his rupture is into right ventricle near the pulmonary valve. Usually this type associated ventricular septal defect (VSD) under the pulmonary valve.
- Type II: aneurysm originated in the middle portion of the RCS and his rupture is into right ventricle. VSD is uncommon.
- Type III: aneurysm has its origin in the middle portion of the right coronary leaflet protruding toward the tricuspid valve. The rupture of this type is into right atrium and, sometimes, in right ventricle under the tricuspid valve insertion.
- Type IV: aneurysm originated in the right portion of the non-coronary leaflet and ruptures into the right atrium (RA). The clinical picture can sometimes mimic pulmonary thromboembolism that can be a catastrophic confusion when a unnecessary thrombolytic therapy is administrated.

CASE REPORT

We present the case of a male patient, 28 years old, from rural area, admitted in the Cardiology Clinical, „St. Spiridon” County Hospital Iași in April 2015, without a history of cardiovascular pathology, symptomatic since approximately 1 month, with pain in the right upper quadrant without irradiation, small-to-medium exertional dyspnea, afebrile, dyaphoresis and asthenia which have gradually increased. The clinical examination reveals IV/VI degree (Levine) pluriorifical continuous (“to and fro”) systolic-diastolic murmur, predominantly in the aortic auscultation area, irradiating along the left sternal border, accompanied by a thrill, rounded enlarged liver 3 cm below the rib edge, slightly painful, without declive edema. Should be noted that diastolic accentuation of this “*machinery*” murmur is an important sign of differentiated RSVA from patent ductus arteriosus^{2,14}, missing in this case.

The patient complaints starts just after a sustained physical effort (he was a construction worker) with new onset shortness of breath, fitting a *New York Heart Association* (NYHA) functional class II. The RSVA initial diagnosis was established by echocardiographic exam as outpatient (“Maycor” Medical Center, Galați) and then the patient was sent to our Cardiology Clinic to establish his further therapeutic management.

Here we confirmed (by a new transthoracic echocardiography) the previous diagnosis, objectifying the aorta-right atrium shunt (Figure 1) which significant charge the pulmonary circulation, right heart cavity dilatation and pulmonary hypertension (systolic pulmonary arterial pressure, sPAP ~50 mmHg), left atrium diameters 49/63/44 mm, ascending aorta diameter 30 mm, aortic Vmax 1,6 m/s, Left Ventricular End Diastolic Diameter (LVEDD) 57 mm, Left Ventricular End Systolic Diameter (LVESD) 35 mm, interventricular septum thickness 11 mm, left ventricle walls thickness 11 mm, Left Ventricular Ejection Fraction (LVEF) 67%, SF-38%, RV 34 mm, Right Atrium diameters (RA) 54/52 mm, Right Ventricular End Diastolic Diameter (RVEDD) 21 mm, normal left cardiac cavities, normal segmental and global kinetic, no addition images on valves (Figure 2

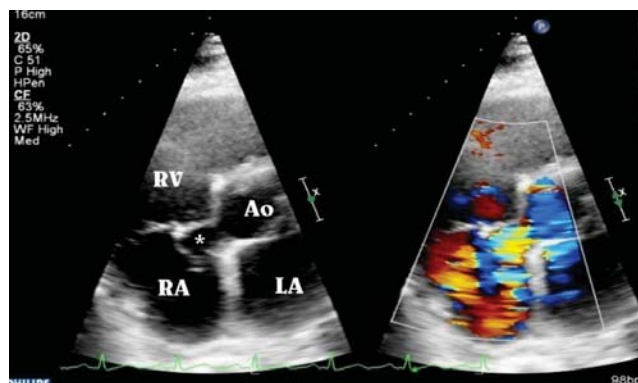


Figure 1. Transthoracic echocardiography (parasternal short axis view, without/with color-flow Doppler mode): aneurysmal dilatation of right sinus of Valsalva (*) protruding into the right atrium (RA); aorta-right atrium shunt with right cardiac chamber (RA and RV) dilatation and severe pulmonary hypertension (systolic pulmonary arterial pressure, sPAP ~ 70 mmHg). Ao aorta; LA left atrium.



Figure 2. Transthoracic echocardiogram (parasternal short axis view): ruptured aneurysm of noncoronary sinus of Valsalva (*) into the right atrium (RA) with left-to-right systolic-diastolic shunt (→) which loads right atrium, right ventricle (RV) and pulmonary circulation. Ao aorta; PA pulmonary artery.

and 3), tricuspid aorta with no vegetations or others valvular abnormalities. Asymmetric dilatation of the aortic root can easily be emphasized (specific feature of “windsock” dilatation). Color flow Doppler revealed a continuous turbulent flow between right sinus of Valsalva and the right atrium through the ruptured aneurysm.

ECG reveals a sinus tachycardia 90 bpm, QRS axis at 45°, normal morphology. A discreet hepatic cytolysis: aspartate aminotransferase (ALAT/TGO) 45/40/40 UI/l, alanine aminotransferase (ALAT/TGP) 148/135/57 UI/l, gamma-glutamyl transpeptidase (GGT) 91/93/112 UI/l, hypoxemia with respiratory alkalosis. Moderate cardiomegaly on chest radiography. Concerning the etiologic diagnosis, it was ruled out syphilis (VDRL negative) and Marfan syndrome (no eyes disorder, normal stature, normal fingers)^{3,4}. Clinical and echocardiographic was also been ruled out other possible malformations associated with ruptured Valsalva aneurysm (aortic insufficiency, aortic stenosis, atrial or ventricular septal defect, coarctation of the aorta). Nor angiography or pulmonary CT angiogram was performed. The patient outcome in our clinic was to haemodynamic rapidly impairment with increasing PASP (70 mmHg) at TTE, low BP, extreme fatigue, increased breathlessness and diaphoresis, symptoms which required the transfer of the patient in surgery ward at Institute of Cardiovascular Diseases from Iași to correct the defect. Treatment for this type of pathology is surgical and consists in closing the communication (the defect correction with Dacron-patch - Figure 4 and 5), after external chest hypothermia, anterograd cardioplegia and right atriotomy approach.



Figure 3. Transthoracic echocardiogram (apical 4-chamber view): ruptured aneurysm of noncoronary sinus of Valsalva into the right atrium which loads right heart circulation.

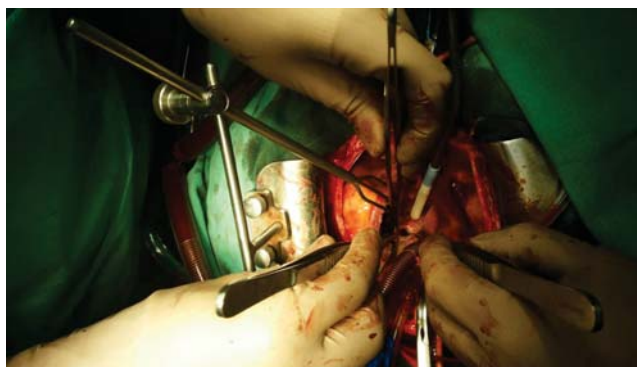


Figure 4. Operative view: noncoronary sinus Valsalva aneurysm ruptured into the right atrium.

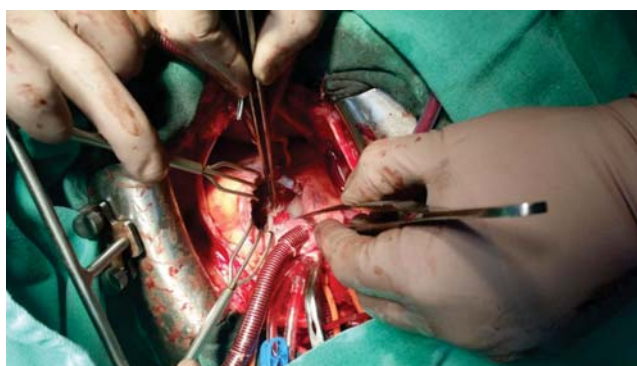


Figure 5. Operative view: correction of rupture with Dacron patch.

Operative results, postoperative course and survival are excellent, he was discharged on eleventh postoperative day with the following recommendations: amlodipine 10 mg once daily, bisoprolol 5 mg once daily, aspirin 75 mg once daily, ranitidine 150 mg once daily. Echocardiography finding at discharge are Left Ventricular End Diastolic Diameter (LVEDD) 55 mm, Left Ventricular End Systolic Diameter (LVESD) 40 mm, Interventricular Septum thickness (IVS) 9 mm, thickness of the left ventricular free wall (LVW) 11 mm, Left Ventricular Ejection Fraction (LVEF) 50%, Left Atrium diameter (LA) 56/50 mm, Right Atrium diameter (RA) 49/40 mm, Right Ventricular End Diastolic Diameter (RVEDD) 36 mm, ascending aorta diameter 37 mm, mitral valve surface area 3.8 cm², Pressure Half Time (PHT aortic) 58 ms, Aortic v_{max} 1,1 m/s, dP_{max} (tricuspid) 20 mmHg, 1st degree mitral regurgitation, 1st degree aortic regurgitation, 1st degree tricuspid regurgitation, LV global systolic function preserved, free heart cavities, pericardium without fluid, at the level of the synthetic patch between right coronary sinus and right atrium it can be seen a very small residual shunt, hemodynamically insignificant. No signs of pulmonary hypertension.

DISCUSSIONS

In young patients (mostly male - 4:1 ratio - and middle-aged), the ruptured aneurysms of sinus of Valsalva may initially be asymptomatic or may be manifested by acute heart failure with the emergence of new heart murmurs imposing a differential diagnosis with other conditions that can cause continuous murmur such as patent ductus arteriosus, aorto-pulmonary window, coronary fistula, ventricular septal defect and aortic regurgitation. SVA without rupture can cause obstruction of the right ventricular outflow tract and may be complicated by endocarditis^{11,18}, thrombus formation, embolic events, myocardial infarction (compression of the coronary artery branches by the aneurysm), cardiogenic shock and death²¹. The symptoms described in patients with unruptured SVA is dominated by medium-small effort dyspnea, palpitations and angina. In addition to the echocardiography, the angiographic study is used to accurately detect the site of the rupture, as well as to estimate the permeability of the coronary vessels. The median diameter of lesions was 6 mm (4-8 mm)²². MRI can be used to confirm the diagnosis^{23,24}, aneurysmal sac position, direction of expansion, the heart cavity in which rupture occurred and other coexisting injuries.

Approximately a third of patients with RSVA with left-right shunt complain angina, breathless and fatigue, symptoms that worsen progressively requiring further investigation. Electrocardiogram can objectify left/right ventricle hypertrophy and non-specific ST segment/T wave abnormalities.

The management of RSVA is guided by clinical and echocardiographical findings. Transesophageal echocardiography is a further examination which can provide valuable data as compared to with transthoracic echocardiography for the aneurysm and coexisting cardiac malformations description. Due to the coexistence of two systolic jets in the right atrium, the difficulty of measuring PAPs should be mentioned. In fact, in our case report the disappearance of pulmonary arterial hypertension after surgical repair indicated that the component responsive to elevated pre-operative sPAP values was the aortic jet. Additional diagnosis methods include cardiac catheterization, computed tomography angiography and cardiovascular magnetic resonance¹⁰. Early diagnosis is important for emergency surgery.

Vural et al¹⁹ used a simple and functional algorithm of the therapeutic approach to ASVs, based on the clinical picture and the echocardiographic findings: endocardial ruptured (type A) or unruptured (asym-

ptomatic - type B-I / symptomatic - type B-II) and extra-cardiac (type C).

Surgical approach is required for both ruptured/unruptured (but symptomatic) aneurysms. SVA become symptomatic when their size exceeds 50% of the average size of the other two sinuses and when there is a compression of the surrounding tissues^{25,26}. The surgical treatment have a low risk and has a long-term survival rate similar to that of healthy individuals when not coexist other malformations (in a series of 19 cases enrolled between 2009-2013 the hospital mortality rate was 5.2%)²⁷. Surgical correction can be summarized in a simple suture or graft plasty with patch closure of the defect²⁷. Because of RSVA rarity there have been no clinical data to support one or other closure technique (primary vs patch closure) and preferred surgical approach (dual, transaortic, involved chamber), but using a patch, as in our reported case, minimize aortic leaflet distortion and reduces the stress on the suture line^{20,27}. The early surgical treatment can prevent the evolution to congestive heart failure, endocarditis or death. In selected RSVA patients percutaneous closure using modified double-disc occluders can be an alternative to surgery²².

The particularity of this case lies in the fact that aneurysm rupture occurred at a young age (28 years) in decade 2-3 of life than the average age cited in the literature (decade 3-4) and without any associated congenital or acquired disease. Our patient have a SVA type III according to the Sakakibara and Konno classification, its origin in the middle portion of the right coronary leaflet, protruding toward the tricuspid valve and the rupture is into right atrium.

CONCLUSIONS

Ruptured sinus Valsalva aneurysms, though rare, can have poor prognosis in the case of delayed diagnosis. An important diagnostic contribution brings the transthoracic echocardiography which confirms the diagnosis and allow the fastest possible surgical treatment without which the patient outcome would be potentially life threatening.

Consent: Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Competing interests: The authors declare that they have no competing interests.

References

1. Chatzis A, Kanakis M, Tsoutsinos A, Mitropoulos F. Ruptured sinus of Valsalva: not always an aneurysm. *Hellenic J Cardiol* 2015; 56: 344-346.
2. Arıkan E, Karagöz A, Bayata S, Yılık L, Ünluer EE. Case report - a rare cause of dyspnea: sudden rupture of aortic Valsalva sinus aneurysm. *Case Reports in Medicine* 2013 <http://dx.doi.org/10.1155/2013/909302>
3. Knight JL, Jacka M, Charrette JP. Repair of isolated, symptomatic sinus of Valsalva aneurysm in a patient with Marfan's syndrome. *Can J Cardiol* 1988; 4: 214-216.
4. Chue SH, Hung CR, How SS, et al. Ruptured aneurysm of the sinus of Valsalva in oriental patients. *J Thorac Cardiovasc Surg* 1990; 99: 288-298.
5. Goldberg N, Krasnow N. Sinus of Valsalva Aneurysms, *Clin Cardiol* 1990, 13: 831- 36.
6. Takach TJ, Reul GJ, Duncan JM, Cooley DA, Livesay JJ, Ott DA, Frazier OH. Sinus of Valsalva aneurysm or fistula: management and outcome. *Ann Thorac Surg* 1999; 68: 1573-7.
7. Smith WA. Aneurysm of the sinus of Valsalva, with report of 2 cases. *J Am Med Assoc* 1914; 62: 1878.
8. Shah RP, Ding ZP, Ng AS, Quek SS. A ten-year review of ruptured sinus of Valsalva: clinico-pathological and echo-Doppler features. *Singapore Med J* 2001; 42: 473-6.
9. Felsoci M, Svatopluk N, Andrasina T, Nemeč P. Ruptured aneurysm of non-coronary sinus of Valsalva as a rare cause of chest pain. *Eur Heart J* 2014; 35(31): 2123.
10. Abdelsalam M, Bachinsky W, Pawlusch D, Mumtaz M, Goldman J. Non-coronary sinus of Valsalva aneurysm rupture into right atrium. *Tex Heart Inst J* 2013; 40: 493-494.
11. Shumacker Jr HB. Aneurysms of the aortic sinuses of Valsalva due to bacterial endocarditis, with special reference to their operative management. *J Thorac Cardiovasc Surg* 1972; 63: 896-902.
12. Wang ZJ, Zou CW, Li DC, et al. Surgical repair of sinus of Valsalva aneurysm in Asian patients. *Ann Thorac Surg* 2007; 84: 156-60.
13. Lakoumentas JA, Bonou MS, Brili S, Theocharis CS, Benroubis AD, Perpinia AS, Harbis PK. Ruptured aneurysm of the right sinus of Valsalva into the right ventricle. *Hellenic J Cardiol* 2002; 43: 242-245.
14. Chang C-C, Chin C-H, Chen M-L, Chen T-H, Lo H-S. Sinus of Valsalva Aneurysm with Rupturing into the Right Atrium - A Case Report and Review of the Literature. *Acta Cardiol Sin* 2006; 22: 96-101.
15. Khandeparkar JM, Porwal MM, Agarwal ChU, Mahajan VD. Bicameral rupture of aneurysm of sinus of Valsalva. *Ind J Thorac Cardiovasc Surg* 2009; 25: 198-200.
16. Van Son J, Danielson GK, Schaff HV, Orszulak TA, Edwards WD, Seward JB. Long-term outcome of surgical repair of ruptured sinus of Valsalva aneurysm. *Circulation* 1994; 90: 1120-1129.
17. Koh KK, Lee KH, Kim SS, Lee SC, Jin SH, Cho SW. Ruptured aneurysm of the sinus of Valsalva in a patient with Behcet's disease. *Int J Cardiol* 1994; 47: 177-179.
18. Biabnam KR, Roberts WC. Fatal intrapericardial rupture of sinus of Valsalva aneurysm. *Am Heart J* 1990; 120(Pt 1):1455-1456.
19. Vural KM, Sener E, Tasdemir O, Bayazit K. Approach to sinus of Valsalva aneurysms: a review of 53 cases. *Eur J Cardio-thorac Surgery* 2001; 20: 71-76.
20. Sakakibara S, Konno S. Congenital aneurysm of the sinus of Valsalva: anatomy and classification. *Am Heart J* 1962; 63: 402-424.
21. Gupta S, Chitlangia R, Garg D. Echocardiographic diagnosis of ruptured sinus of Valsalva with congenital VSD and AR. *J Clin Prev Cardiol* 2015; 4: 24-6.
22. Liu S, Xu X, Zhao X, Chen F, Bai Y, Li W, Zhang Y, Wang C, Xiang J, Wu G, Chen X, Qin Y. Percutaneous closure of ruptured sinus of Valsalva aneurysms: results from a multicentre experience. *EuroIntervention* 2014; 10: 505-12.
23. Dev V, Goswami KC, Shrivastava S, Bahl VK, Saxena A. Echocardiographic diagnosis of aneurysm of the sinus of Valsalva. *Am Heart J* 1993; 126: 930-936.
24. Yılmaz AT, Demirkılıç U, Oza E, et al. Aneurysm of the sinus of Valsalva. *J Cardiovasc Surg* 1997; 38: 119-124.
25. Mayer ED, Ruffmann K, Saggau W, Butzmann B, Mayer KB, Schatton N, Schmitz W: Ruptured aneurysms of the sinus of Valsalva. *Ann Thorac Surg* 1986; 42: 81-85.
26. Tami LF, Turi ZG, Arbulu A. Sinus of Valsalva aneurysms involving both coronary ostia. *Cath Cardiovasc Diag* 1993; 29:304-308.
27. Yadav A, Mathur R, Devgarha S, Abraham V, Sisodia A. Surgery for ruptured sinus of Valsalva aneurysm: Five-year experience with 19 patients. *Turk Gogus Kalp Dama* 2014; 22: 729-733.