

# Sinus of Valsalva aneurysm with rupture. Case report and literature review

Matilde Myriam Galicia-Tornell,\* Bertha Marín-Solís,\*\* Oscar Mercado-Astorga,\*  
Saúl Espinoza-Anguiano,\*\* Manuel Martínez-Martínez,\* and Esteban Villalpando-Mendoza\*

## Abstract

**Background:** The sinus of Valsalva aneurysm (SVA) is a small dilatation caused by a separation between the aortic media and annulus fibrosus. Its origin may be either acquired or congenital. The right coronary sinus is most frequently affected, with the most common complication being rupture.

**Clinical cases:** We report two cases of SVA complicated with rupture to the right cavities with severe cardiac failure, which represents 0.4% of the total cardiac surgeries performed during the past year in our hospital.

**Case #1:** We present the case of a 27-year-old male with low-effort dyspnea, nocturnal paroxysmal dyspnea, orthopnea, paroxysmal atrial fibrillation, diastolic murmur, hepatomegaly and edema of the lower extremities.

**Case #2:** We present the case of a 33-year-old male with low-effort dyspnea, nocturnal paroxysmal dyspnea, orthopnea, diastolic murmur, severe kidney insufficiency and congestive hepatic insufficiency.

Surgical resolution in both cases consisted of aneurysm resection and closure of the defect with a Dacron patch, preserving the aortic valve. Both patients survived.

**Conclusions:** Cardiac failure and sudden auscultation changes suggest the possibility of SVA rupture. Echocardiography is sufficient to diagnose SVA, its complications, repercussions, and surgical options. SVA, even if asymptomatic, has potential risks of expansion, rupture, cardiac failure, endocarditis, embolism and sudden death. This justifies surgical correction, with a low mortality rate in both the short- and long-term.

**Key words:** Sinus of Valsalva, aneurysm rupture.

## Introduction

Sinus of Valsalva (SV) is a dilatation of the aortic wall located between the aortic valve and the sinotubular junction. Its location is related to the coronary arteries designated as the right coronary sinus (RCS), left coronary sinus (LCS) and noncoronary sinus (NOCS).<sup>1-3</sup> SVA is a dilatation caused by the lack of continuity between the middle layer of the aortic wall and the aortic valve.<sup>1,2,4,5</sup> It is a rare disease that has been reported in 0.09% in

one autopsy series, but in Western surgical studies incidence of 0.14-0.23% has been reported and 0.46-3.5% in an Asian series.<sup>1,3-5</sup> The most common cause is congenital, although its origin may be acquired (trauma, infection, or degenerative diseases).<sup>2-5</sup> It commonly coexists with other malformations such as ventricular septal defect (VSD), anomalies of the aortic valve and coarctation of the aorta.<sup>1,3,4</sup> RCS is the one most frequently affected followed by the NOCS and, rarely, the LCS.<sup>1-4,6</sup> SVA are asymptomatic unless they compress adjacent structures during their expansion or form a thrombus.<sup>1,4</sup> The most common complication is rupture into the atrium or ventricle, very rarely towards the left chambers, causing left-to-right shunting or aortic valve insufficiency with congestive heart failure and the need for urgent surgical resolution.<sup>1-4,6</sup> Unruptured SVA are undetected lesions because they are asymptomatic, suggesting continuous monitoring and indicating surgery only if they demonstrate aneurysm expansion, rupture or infection. However, the potential risk of rupture, cardiac insufficiency (CI), stroke and sudden death has led other authors to consider surgical repair of unruptured aneurysms even if they are asymptomatic or incidentally detected, reporting a generally low surgical mortality.<sup>1,2,4,6</sup> In our hospital, correction of SVA corresponds to 0.4% of the cardiac surgeries performed during 1 year.

\* Servicio de Cirugía Cardioráfrica.

\*\* Servicio de Cardiología.

Hospital de Especialidades, Centro Médico Nacional de Occidente, Instituto Mexicano del Seguro Social, Guadalajara, Jalisco, Mexico

Correspondence and reprint requests to:

Matilde Myriam Galicia-Tornell

Bosques de Birmania 27-B101

Col. Bosques de Aragón

57170 Nezahualcóyotl, Estado de México

Tel: (55) 5780 1123; (55) 5796 8573

E-mail: mmygato@hotmail.com

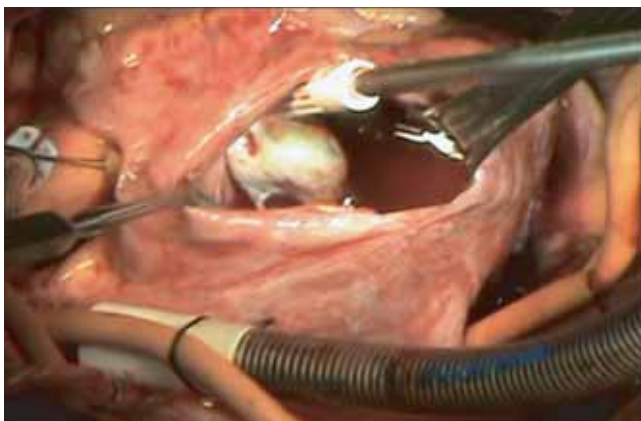
Received for publication: 4-13-2009

Accepted for publication: 8-3-2009

We report on two patients with aneurysms of the right SVA and unruptured coronaries in the right cavities, causing CI. These were surgically resolved, illustrating the importance of opportune diagnosis and treatment, as well as the need for long-term monitoring.

### Clinical Case 1

We present the case of a 27-year-old male with a history of heart murmur and hospitalizations for CI, which progressed to low-effort paroxysmal nocturnal dyspnea, orthopnea, edema of the lower extremities and palpitations. He presented clinically with arrhythmic heart sounds, diastolic murmur GIII/VI, hepatomegaly and edema of lower extremities. Chest x-ray showed grade IV cardiomegaly. We reported an EKG with paroxysmal atrial fibrillation. Transthoracic echocardiogram (TTE) showed right ventricle (RV) with severe dilatation and mild systolic dysfunction, mild tricuspid insufficiency, severe biatrial dilatation predominantly on the right, noncoronary aneurysm SVA ruptured in the right atrium (RA), type IV classification of Sakakibara and Konno. Three-valved aortic valve had no insufficiency or VSD. Corrective surgery was performed where grade IV heart enlargement was observed at the expense of the right chambers, severely dilated RA, normal aortic valve with noncoronary SVA of 2.5 cm in length, ruptured towards the RA, and adhered to the tricuspid valve without affecting its functionality (Figure 1). It contained a sinus rhythm. The repair was performed with the assistance of a cardiopulmonary bypass (CPB) and moderate hypothermia (32°C). The approach was made through aortotomy and right atriotomy. The aneurysm was exposed and resected from the RA and the defect was closed with a Dacron patch with a prolene suture 4-00 (Figure 2). CPB was 83 min and aortic clamping lasted 55 min. The patient's postoperative evolution was uncomplicated. Histopathological examination showed segments with wall thinning and with



**Figure 1.** Rupture of noncoronary sinus of Valsalva aneurysm (SVA) into right atrium.

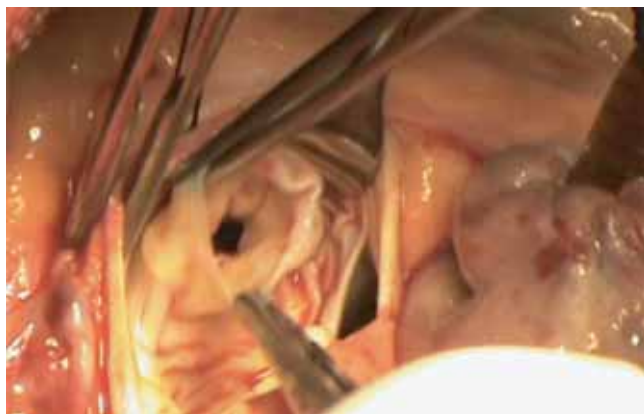
collagen fibers replacing smooth muscle cells. Intraoperative echocardiogram showed a residual leak with mild aortic valvular insufficiency.

### Clinical Case 2

We present the case of a 33-year-old male patient. He was an athlete and had a history of unspecified congenital heart disease, without surgery. He presented with CI for 4 months, which included a hospital stay. He persisted with low-effort dyspnea, orthopnea and paroxysmal nocturnal dyspnea. Clinical report included jaundice, grade IV jugular plethora, frequent extrasystoles, continuous murmur of grade III/VI, crackling rales, hepatomegaly, wide peripheral pulses, and edema of the lower extremities. We documented acute renal failure and congestive liver failure. Electrocardiogram showed a sinus rhythm, incomplete blockage of right bundle branch of His, and frequent and multiform ventricular extrasystoles. Chest x-ray showed a grade III cardiomegaly at the expense of the RV and pulmonary hyperflow. TTE showed left ventricular hypertrophy and normal systolic function; RV showed dilatation and severe hypertrophy and moderate systolic dysfunction without IVSD. Right SVA showed rupture and 17-mm protrusion for RV, type II classification of Sakakibara and Konno, and left-to-right shunting. Three-valved aortic valve had a mild impairment and mild tricuspid insufficiency. There was severe dilation of the RA. The patient underwent corrective surgery, confirming a right SVA (2 cm long) and ruptured RV (Figure 3). The repair was performed with the assistance of CPB and moderate hypothermia (32°C). Approach was made through an aortotomy. The aneurysm was exposed and resected from the aorta (Figure 4). The defect was closed with a Dacron patch with continuous suture and separate stitches with prolene 4-00. Competency was confirmed through direct vision of the aortic valve (Figure 5). CPB time was 90 min and time for aortic clamping was 73 min. Intraoperative echocardiogram discarded residual and



**Figure 2.** Resection of aneurysmatic sac of noncoronary SV.



**Figure 3.** Right SVA rupture into right ventricle.



**Figure 4.** Aortic exposure and resection of right SVA.



**Figure 5.** Closure of the defect with Dacron patch and comprobation of aortic valve competency.

insufficient valvular leakage. No complications were reported during his hospital stay. Histopathological study reported thinned segments with replacement of smooth muscle cells by collagen without elastic fibers. There was no inflammation, necrosis or any microorganisms.

## Discussion

SVAs have a low frequency; 0.15-1.5% of cardiac surgeries correspond to SVA repair.<sup>1,4-13</sup> This range is attributed to ethnic variation. Chu et al. reported that the incidence of ruptured SVA is five times higher in Asian countries (0.46-3.5% in Eastern and 0.14-0.23% in Western areas).<sup>3,7-9,11,13,14</sup> Of the group, 65-80% are males (male/female ratio 4:1).<sup>3,6,8,15</sup> Diagnosis can occur at any age (average: 39 years old, range: 2-74 years).<sup>1,2,3,8,11,14</sup> SVA affect the right sinus in 65-86% of cases, noncoronary sinus in 10-30% and the left sinus in 2-5%.<sup>1-4,6-8,10,11,14,15</sup> When aneurysmal dilatation involves the left sinus or there are multiple SV of uncertain congenital origin, it is considered an aneurysmal dilatation of the

aortic root with different clinical presentations and surgical treatment.<sup>3,6,10,14</sup> SVA can be congenital or have an acquired origin, which may be due to infection, trauma or degenerative diseases.<sup>2-5</sup> They are associated with other heart defects, VSD in 30-60% of cases, aortic valve abnormalities such as aortic insufficiency in 20-30% of cases, bicuspid valve (10%), aortic stenosis (6.5%). They may also be associated with pulmonary stenosis (9.7%), coarctation of the aorta (6.5%), persistence of the conductus arteriosus (3.2%), tricuspid insufficiency (3.2%) and interatrial defect.<sup>1,7,9-11,13-15</sup> The frequency of ruptured SVA varies according to the location: 60% in the right sinus, 42% in the noncoronary sinus and only 10% in the left sinus. Rupture may be spontaneous, after trauma, extreme physical exercise or due to endocarditis.<sup>3</sup> Rupture of a SVA occurs principally at the RV (60%) or at the RA (29%), the LA (6%), LV (4%) or at the pericardium (1%). Extracardiac ruptures are rare, usually fatal, and occur towards the pericardium or the pleural space and are more common when the SVA is of acquired origin.<sup>1,2,4,7,8,11,13,14</sup> Physiopathological consequences of SVA rupture depend on the volume of flow through communication, velocity of establishment of the rupture and cardiac chamber with which it communicates. The perforation of acutely high flow does not allow hemodynamic compensation, developing a sudden CI.<sup>2,6,14,15</sup> A small, gradual and progressive perforation can be tolerated temporarily, occurring in 25% of cases.<sup>11</sup> SVA rupture causes thoracic pain, dyspnea, cough, fatigue, tachycardia, or peripheral edema, as well as a continuous murmur. The intact SVA may be asymptomatic, produce thrombus or distortion and compression of the coronary arteries, leading to ischemia.<sup>2,6,10</sup> The only formal classification for the SVA has been proposed by Sakakibara and Konno in 1962, describing four types according to the coronary sinus affected and the area where they protrude or rupture (Table 1).<sup>1-3,15</sup>

EKG findings in the cases of ruptured SVA are ventricular hypertrophy, electrical axis towards the right and atrial fibrillation. In unruptured SVA, EKG is usually normal unless the aneurysm compresses the atrioventricular node or its branches, resulting in

**Table 1.** Classification for SVA proposed by Sakakibara and Konno

Type I	Connect the right SV and the existing tract of the RV below the pulmonary valve
Type II	Connect the right SV and the VD in the <i>supraventricularis</i> crest
Type IIIa	Connect the right SV and the RA
Type IIIv	Connect the posterior zone of the right SV and the RV
Type IIIa+v	Connect the right SV and both RA and RV
Type IV	Connect the noncoronary SV and the RA

SV, sinus of Valsalva; RV, right ventricle; RA, right atrium.

complete blockage or other atrio-ventricular conduction alterations. TTE and transesophageal echocardiogram (TEE) have a diagnostic accuracy of 75% and 90%, respectively, for ruptured or unruptured SVA and allow discrimination of size, origin of the sinus, point of termination, severity and mechanism of the valvular insufficiency, presence of cardiac or associated vascular abnormalities as well as the orientation of the surgical procedure.<sup>1,6,8,10-12,14</sup> Although angiography is considered the gold standard, it is only necessary in some cases. MRI and CT offer high diagnostic effectiveness.<sup>1,6,8,11-13</sup> In 1955, the first successful correction of a SVA was reported under deep hypothermia without CPB.<sup>3</sup> In 1956, CPB was used for the first time for resection of the SVA and primary closure of the defect, as well as the use of cardioplegia.<sup>3</sup> The usual therapeutic option for SVA is surgery. There have been cases published of percutaneous intravascular repair.<sup>1,3</sup> For patients with untreated ruptured SVA, the average survival after diagnosis is 3.9 years, which supports the indication for surgical repair. There is no known history of natural unruptured aneurysms because they are more often asymptomatic. Unruptured SVA that produces malignant arrhythmias, infection, blockage of coronary arteries or ventricular outflow tracts indicate the need for surgery.<sup>1,2,4,6,11</sup> Velocity of the progression of the aneurysm is a factor in the decision because it may be rapid, as reported by Regueiro et al.<sup>4</sup> Therefore, the presence of an unruptured SVA, even if it is asymptomatic or incidentally detected, is an indication for surgical intervention in most cases. An SVA found during heart surgery for another pathology should be repaired, even if it is small.<sup>2,5,10,11</sup> In addition, repair of the asymptomatic SVA has excellent results. The surgical technique depends on the specific anatomy: if the aneurysm is ruptured or if there is an associated heart defect, it is necessary to repair or replace the aortic valve.<sup>4,11</sup> Because of the rarity of SVA, there are no studies that demonstrate which surgical technique is superior. However, the best results were obtained with the aneurysm closure, removal of the aneurysm sac and repair of any other associated defect without causing blockage or dysfunction of the aortic valve.<sup>1,11</sup> In general, surgical repair of SVA is divided into repairs through the chamber of origin (aorta), through the chamber of the penetration or through both chambers. The technique is also classified as primary closure (with or without Teflon reinforcement), aortic root replacement (usually associated with severe aortic insufficiency or compromise of more than one

SV) or patch closure (single or double), which prevents aortic valve deformation, reduces stress on the suture line, and optimizes long-term functionality compared to direct closure.<sup>1</sup> Operative mortality is generally low (1%)<sup>1,8</sup> in patients without infection; an Eastern study registered 3.5% mortality.<sup>7,11,13</sup> However, cases of infected SVA (endocarditis or sepsis) have 4-5 times greater risk of perioperative death. Perioperative mortality in patients without an infection is attributed to low cardiac output, especially in patients with cardiac abnormalities that are corrected concomitantly. Recent series have estimated a survival of 5 and 10 years after the correction of SVA of 97% and 82%, respectively.<sup>1,9</sup> The majority of patients show symptom improvement as well as a decrease in diastolic ventricular diameter.<sup>8</sup> The prognosis seems to be improved if the aortic valve and aortocoronary bridge replacement can be avoided.<sup>4,15</sup> Subsequent events are related to the prosthetic valve (dysfunction, perivalvular flow, thrombosis, endocarditis); aortic pathology basis (Marfan syndrome, aortic dissection) and the need for prosthetic valve implant in cases of repair of the native aortic valve. SVA recurrence per se is rare.<sup>9,15</sup> Our data and that of other hospital centers show the importance of developing a high degree of suspicion of this pathology, as compared to patients who develop sudden CI and abrupt changes in auscultation, considering TTE or TEE sufficient to diagnose SVA. There is no doubt of the indication of surgical repair for symptomatic SVA because this represents a significant risk of death. Operative mortality is generally low and we believe that removal of the aneurysmal sac, closure of the defect with a patch, preservation of the native aortic valve and correction of any concomitant disease are the surgical strategies with the most favorable results and lower risk of recurrence. Long-term follow-up allows the assessment of the events related to functional class, regression of ventricular diameters and function, need for prosthetic valve implant in cases of repair of the native aortic valve, and recurrence.

In conclusion, our cases reflect the expectant behavior for ruptured and unruptured SVA with little hemodynamic repercussion; however, the behavior towards progressive growth of the SVA and the potential risk of rupture, CI, endocarditis, stroke and sudden death all justify surgical resolution of SVA even in asymptomatic cases, considering even better results from uncomplicated SVA.

## References

1. Ott D. Aneurysm of the sinus of Valsalva. *Cardiac Surg Ann* 2006;9:165-176.
2. Caballero J, Arana R, Calle G, Caballero FJ, Sancho M, Piñero C. Aneurisma congénito del seno de Valsalva roto a ventrículo derecho, comunicación interventricular e insuficiencia aórtica. *Rev Esp Cardiol* 1999;52:635-638.
3. Steves W. Congenital heart surgery nomenclature and database project: aortic aneurysm, sinus of valsalva aneurysm, and aortic dissection. *Ann Thoracic Surg* 2000;69:S147-S163.
4. Regueiro M, Penas M, López V, Castro A. Aneurisma del seno de Valsalva como causa de un infarto agudo de miocardio. *Rev Esp Cardiol* 2002;55:77-79.
5. Vautrin E, Barone-Rochette G, Philippe J. Rupture of right sinus of Valsalva into right atrium: ultrasound, magnetic resonance, angiography and surgical imaging. *Arch Cardiovasc Dis* 2008;101:501-502.
6. Missault L, Callens B, Taeymans Y. Echocardiography of sinus of Valsalva aneurysm with rupture into the right atrium. *Int J Cardiol* 1995;47:269-272.
7. Chu SH, Hung CR, How SS, Chang H, SS Wang, Tsai CH, et al. Ruptured aneurysms of the sinus of Valsalva in Oriental patients. *J Thorac Cardiovasc Surg* 1990;99:288-298.
8. Dong C, YuWu Q, Tang Y. Ruptured sinus of Valsalva aneurysm: a Beijing experience. *Ann Thorac Surg* 2002;74:1621-1624.
9. Harkness J, Fitton T, Barreiro CJ, Alejo D, Gott VL, Baumgartner WA. A 32-year experience with surgical repair of sinus of Valsalva aneurysms. *J Cardiac Surg* 2005;20:198-204.
10. Vázquez C, Erdmenger J, Roldán FJ, Romero A, Vargas J. Valoración ecocardiográfica bi y tridimensional de aneurisma no roto del seno de Valsalva. *Arch Cardiol Mex* 2005;75:148-153.
11. Leos A, Benavides M, Nacoud A, Rendón F. Aneurisma del seno de Valsalva con rotura al ventrículo derecho, relacionado con comunicación interventricular perimembranosa. *Med Univ* 2007;9:77-81.
12. Marciani G, Pulita M, Boccio E, Verdugo RA. Rotura de aneurisma del seno de Valsalva coronario derecho. A propósito de un caso. *Rev Fed Arg Cardiol* 2006;35:186-188.
13. Sánchez M, García M, Quintana C, Kareh J. Heart failure in rupture of a sinus of valsalva aneurysm. *Am J Med Sci* 2006;331:100-102.
14. Lobato E, Greene M, Malias M, Gravenstein N. Intraoperative usefulness of transesophageal echocardiography: detection of unsuspected rupture of an aneurysm of the sinus of Valsalva. *J Cardiothorac Vasc Anesth* 1997;11:619-621.
15. Beck M, Blanco F, Márquez E, Papantoniou A, Spornanzoni F, Lax J, et al. Aneurisma congénito del seno de Valsalva roto a ventrículo derecho asociado a comunicación interventricular. *Rev CONAREC* 2003;71:158-160.