Sudden heart failure due to a ruptured posterior Valsalva sinus aneurysm into the right atrium: feasibility of catheter closure using the Amplatzer duct occluder

Eustaquio Onorato, Francesco Casilli, Marcel Mbala-Mukendi, Elena Perlasca, Franco Santoro, Franco Bortone*, Vincenzo Arena**

*Division of Cardiology, *Division of Anesthesiology, **Division of Cardiac Surgery, Humanitas Gavazzeni Clinic, Bergamo, Italy*

Key words:
Aneurysm; Congenital heart disease; Heart failure; Interventional procedures; Intravascular ultrasound.

Valsalva sinus aneurysms are usually congenital and relatively rare and tend to be more frequent in adults. Rupture of these aneurysms can result in sudden death or in an abrupt and rapid progressive heart failure. Surgical repair is the traditional treatment of choice. We report the case of a 48-year-old female with a ruptured posterior non-coronary Valsalva sinus aneurysm, resulting in an anomalous aorto-right atrial fistula. Successful percutaneous catheter closure of the massive left-to-right shunt by using the Amplatzer duct occluder is presented.

(Ital Heart J 2005; 6 (7): 603-607)

Received December 14, 2004; revision received February 25, 2005; accepted February 28, 2005.

Address:
Dr. Eustaquio Onorato
U.O. di Cardiologia I
Clinica Humanitas Gavazzeni
Via M. Gavazzeni, 21
24125 Bergamo
E-mail:
eonorato@libero.it

Introduction

The Valsalva sinus aneurysms are usually congenital and relatively rare. This abnormality seems to be more common in Oriental patients[1].

Sudden rupture of these aneurysms causes a dramatic clinical presentation with abrupt and rapid progressive heart failure.

Since the late '50s, the surgical patch closure of the fistula with the excision of the aneurysm and the treatment of any associated aortic valve abnormalities[2-4] by means of cardiopulmonary bypass has been the standard option.

Only a few cases of percutaneous device closure of ruptured Valsalva sinus aneurysms have been published in the literature[5-8].

We report a case of a ruptured non-coronary Valsalva sinus into the right atrium which has been treated percutaneously by means of an Amplatzer duct occluder (ADO).

Case report

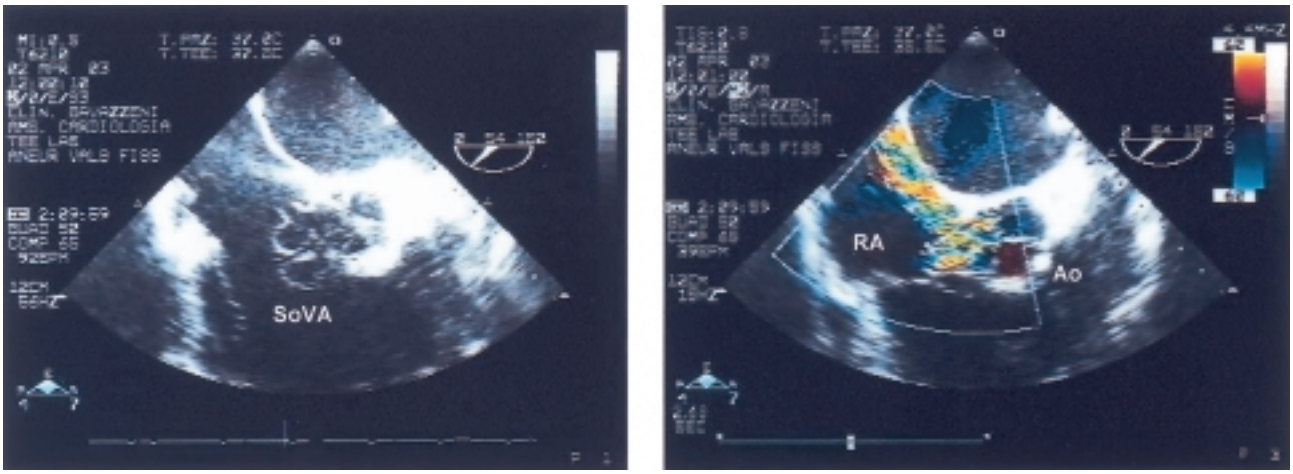
A 48-year-old female with thalassemia trait presented to our Cardiovascular Center with new-onset dyspnea and progressive congestive heart failure of unknown origin. She was known to have borderline systemic hypertension.

Her cardiac examination revealed the presence of a continuous murmur on the left sternal border. An ECG displayed sinus tachycardia and incomplete right bundle branch block without arrhythmias or ST-segment changes.

Chest X-ray showed a right pleural effusion and mild cardiomegaly. Transthoracic echocardiography revealed a normal left ventricular function and size with mild postero-lateral pericardial effusion. The right atrium was slightly enlarged. Furthermore, there was an abnormal circular thin-walled structure posterior to the aortic root, protruding into the right atrium, with a significant left-to-right shunting from the aorta to the right atrium and with a peak velocity of 6 m/s.

A transesophageal echocardiographic study confirmed the presence of a ruptured Valsalva sinus aneurysm originating from the posterior non-coronary sinus (type IV Sakakibara classification)[9] into the right atrium. The color jet was directed towards the atrial septum (Fig. 1).

In the cardiac catheterization laboratory her pulmonary artery pressure was 50/30 mmHg (mean 36 mmHg) with a QP:QS ratio of 4:1. A selective coronary angiography showed a normal coronary artery anatomy. A three-leaflet aortic valve without insufficiency and a ruptured non-coronary Valsalva sinus aneurysm into the right



A B

Figure 1. A: *transesophageal echocardiographic evidence of the presence of a ruptured Valsalva sinus aneurysm (SoVA) originating from the posterior non-coronary sinus of the aortic valve (Ao) into the right atrium (RA);* B: *the color jet is directed towards the atrial septum.*

atrial chamber were detected at ascending aortography (Fig. 2). The procedure was interrupted for further discussion with the patient and her family about the type of management.

After a careful discussion about surgical vs device closure, the patient was sent to the catheterization laboratory a few days later. Under local anesthesia, arterial and venous femoral access were obtained and a total of 7500 IU heparin was administered. The procedure was monitored by transthoracic echocardiography and intracardiac echocardiography (ICE) using a 9F, 9 MHz Ultra-ICE catheter (EP Technologies, Boston Scientific Corporation, San Josè, CA, USA). In order to obtain two standardized orthogonal planes (the transverse aortic valve and the longitudinal four-chamber planes) the ICE catheter was positioned in the right atrium, providing a 360° radial imaging plane perpendicular to the long axis of the catheter as previously described in the literature[10,11]. In this particular case, the ICE imaging modality allows us to visualize a precise relationship between the Valsalva sinus aneurysm and the closer cardiac structures (aortic and tricuspid valves) during the entire procedure.

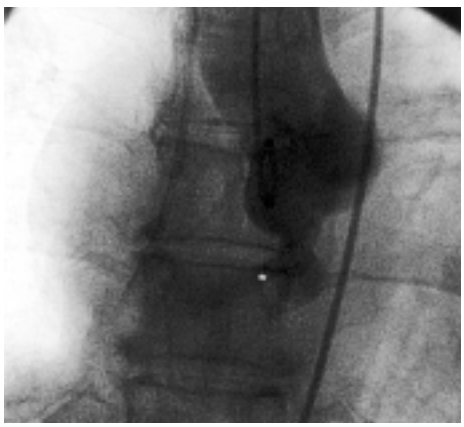


Figure 2. *Ascending aortography showing a normal aortic valve without insufficiency and the presence of a ruptured non-coronary Valsalva sinus aneurysm into the right atrial chamber (*).*

A multipurpose catheter was advanced retrogradely from the femoral artery through the ruptured non-coronary Valsalva sinus into the right atrial chamber and subsequently maneuvered to the inferior vena cava.

An Amplatz Super Stiff 0.035" 260 cm long guidewire was then advanced via the multipurpose catheter to the inferior vena cava, snared and exteriorized out the right femoral vein thus creating an artero-venous wire loop. Subsequently, a 6F Mullins type delivery sheath (AGA Medical Corporation, Golden Valley, MN, USA) was advanced into the ruptured aneurysm through the loop.

Measurements of the fistula diameter were obtained mainly by means of angiography as usually performed during catheter closure of the patent ductus arteriosus (device size at least 2 mm larger than the narrowest portion of the fistula). Moreover, a sizing balloon maneuver was not performed in order to avoid any risk of tissue damage and potential increase in rupture.

A first attempt to place an 8/6 mm ADO was unsuccessful due to a premature and unexpected unscrewing of the device which finally migrated to a left secondary pulmonary artery branch without compromising blood flow. A percutaneous attempt to retrieve the embolized device was ineffective.

A new artero-venous wire loop was formed and a new 7F Mullins type delivery sheath was advanced from the femoral vein into the aneurysm (Fig. 3). A 10/8 mm ADO was then successfully deployed across the site of Valsalva sinus rupture (Fig. 4) and the appropriate placement of the device was confirmed both at transthoracic echocardiography and ICE (Fig. 5).

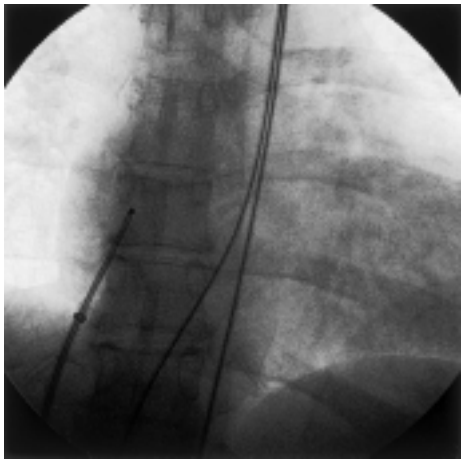


Figure 3. *Artero-venous wire loop across Valsalva sinus aneurysms.*

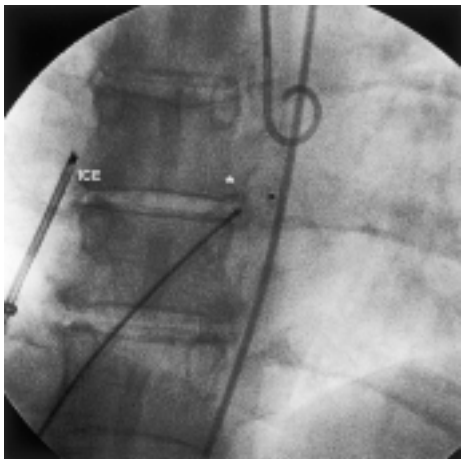
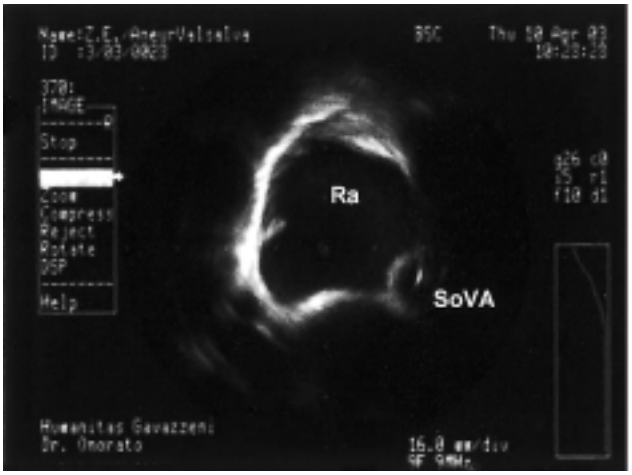


Figure 4. *A 10/8 mm Amplatzer duct occluder (*) deployed across the site of rupture of the Valsalva sinus and intracardiac echocardiography (ICE) catheter positioning in the right atrium.*



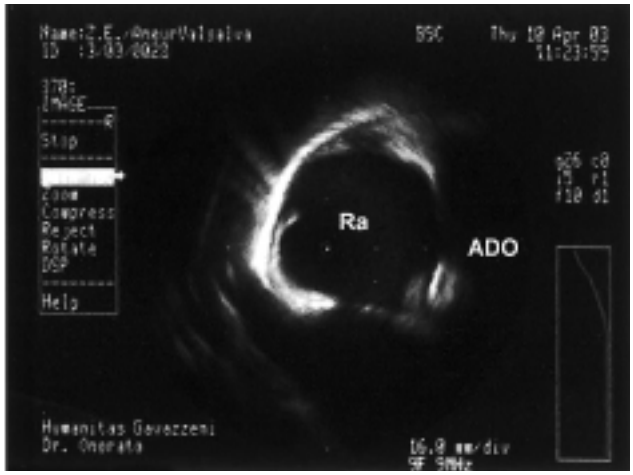


Figure 5. *Intracardiac echocardiography confirming the appropriate placement of the device without impingement on the tricuspid valve apparatus. ADO = Amplatzer duct occluder; Ra = right atrium; SoVA = Valsalva sinus aneurysm.*

A repeat ascending aortography revealed a complete closure of the rupture site (Fig. 6) without aortic insufficiency, and the hemodynamic assessment revealed that the patient's QP:QS ratio was 1:1. Fluoroscopy time was 60 min. Endocarditis antibiotic prophylaxis (ciprofloxacin 200 mg i.v.) was administered during the procedure.

The day later, a transthoracic echocardiography revealed normal left and right ventricular function and size. There was no aortic insufficiency and no residual shunt. The device position was stable. The continuous murmur disappeared and the patient was discharged on 100 mg/day aspirin for 6 months.

No symptoms or adverse events were observed during follow-up and echocardiographic evaluation performed at 3, 6 and 12 months showed no evidence of significant residual shunt.

A thoracic computed tomography scan performed 12 months later revealed a stable position of the migrated ADO in the left secondary pulmonary artery without evidence of thrombosis or pulmonary infarction.

Figure 6. *Final ascending aortography showing complete closure of the site of rupture with no aortic insufficiency.*

Discussion

Valsalva sinus aneurysms are usually congenital and relatively rare, accounting for 0.1-3.5% of all congenital heart disease with a male predominance[12]. The incidence appears to be higher in the Oriental populations[1]. The aneurysm arises from incomplete fusion of the distal bulbar septum that divides the pulmonary artery and the aorta and attaches to the annulus fibrosus of the aortic valve[13]. There is also a relative deficiency of elastic fibers in the affected sinus that, after exposure to long-standing high pressures, progressively dilates over time. Most series have shown that Valsalva sinus aneurysms arise with a strikingly high prevalence from the right sinus of Valsalva (75-90%), whereas the non-coronary sinus is affected in 5-15% of patients. On the contrary, the left sinus is not derived embriologically from bulbar septum and therefore it is rarely affected by congenital lesions.

Congenital Valsalva sinus aneurysms are often isolated but an association with ventricular septal defect, aortic insufficiency and persistent left superior vena cava has been reported[14,15].

Acquired Valsalva sinus aneurysms are less common, as the left sinus is more frequently involved. Secondary causes include bacterial endocarditis, tuberculosis, syphilis, trauma, degenerative and inflammatory processes[14-17].

If untreated, Valsalva sinus aneurysms may remain clinically silent for many years. The only risk is bacterial endocarditis and progressive heart failure if rupture occurs. Symptoms are sometimes caused by mechanical compression of the conducting system leading to heart block[17] or by extrinsic narrowing of the coronary arteries, leading to an unusual cause of myocardial ischemia.

The rupture of Valsalva sinus aneurysms occurs commonly in the third or fourth decade of life with a clinical scenario consisting of dyspnea, chest pain and heart failure in a patient with a recent continuous murmur. The rupture results in a fistulous communication between the sinus of Valsalva and the right heart chambers, namely the right ventricle. If the rupture arises from the non-coronary sinus of Valsalva, the right atrium is more frequently involved.

Chest X-ray does not help in the diagnosis of unruptured Valsalva sinus aneurysms. However, an enlarged heart or pulmonary venous congestion may be demonstrated if rupture or aortic insufficiency occur.

In the past, angiography[18] was the diagnostic tool of choice. However, transthoracic color Doppler echocardiography can delineate the exact location of the rupture and is considered to be the imaging technique of choice[19-21].

ICE is the newest imaging modality and it is a relatively new technique that is still evolving.

On the basis of our experience regarding ICE evaluation during catheter-based closure of atrial septal defect and patent foramen ovale[10,11,22], to the best of our knowledge, this is the first case of a ruptured Valsalva sinus aneurysm closure using ICE for diagnostic and monitoring purposes. This minimally invasive, new imaging modality was very useful in delineating the wall of the aneurysm as a circular thin-walled structure.

Until recently, the treatment of choice was the repair by means of cardiopulmonary bypass with an acceptable low operative risk and a good long-term outcome[23-26]. Operative mortality is < 2%. A systematic approach for the timing of Valsalva sinus aneurysm repair has been proposed by Vural et al.[27].

Percutaneous treatment of most left-to-right shunting lesions has become possible thanks to the recent technological advances.

The first description of such a percutaneous closure of a ruptured Valsalva sinus aneurysm using a Rashkind umbrella was reported by Cullen et al.[5]; more recently, the use of ADO for closing a rupture of the right Valsalva sinus aneurysm into the right ventricular outflow tract has been reported[6].

Rao et al.[7] reported on a transcatheter occlusion of a ruptured Valsalva sinus aneurysm using the bioptome-assisted delivery of a Gianturco coil.

The use of ADO to repair ruptured Valsalva sinus aneurysm has been recently reported by Fedson et al.[8]. Although designed for patent ductus arteriosus and vascular fistula closure, this occluder device achieved a complete closure of the two sites of rupture.

Our case extends the use of ICE imaging during percutaneous interventional procedure; in fact, this tool could replace transesophageal echocardiographic monitoring during the procedure, thus abolishing the need for general anesthesia without additional patient discomfort. However, the major drawback of the Ultra-ICE is still the lack of color Doppler advantages.

In conclusion, this case outlines the efficacy and safety of non-surgical closure of ruptured Valsalva sinus aneurysm. Furthermore, the innovative use of ICE guidance during closure adds this tool to the armamentarium of imaging modalities during transcatheter interventional procedures.

References

1. Chu SH, Hung CR, How SS, et al. Ruptured aneurysms of the sinus of Valsalva in Oriental patients. J Thorac Cardiovasc Surg 1990; 99: 288-98.
2. Kirklin JW, Barratt-Boyes BG. Congenital aneurysm of sinus of Valsalva. In: Kirklin JW, Barratt-Boyes BG, eds. Cardiac surgery. 2nd edition. New York, NY: John Wiley and Sons, 1993: 825.
3. Hoffman JI. Congenital anomalies of the coronary vessels and the aortic root. In: Emmanouilides GC, Riemenschneider TA, Allen HD, Gutdesell HP, eds. Moss and Adams heart disease in infants, children and adolescents. 5th edition. Baltimore, MD: Williams and Wilkins, 1998: 679.
4. Hamid IA, Jyothi M, Rajan S. Transaortic repair of ruptured

aneurysm of sinus of Valsalva: fifteen-year experience. J Thorac Cardiovasc Surg 1994; 107: 1464-8.
5. Cullen S, Somerville J, Redington A. Transcatheter closure of a ruptured aneurysm of the sinus of Valsalva. Br Heart J 1994; 71: 479-80.
6. Cullen S, Vogel M, Deanfield JE, Redington AN. Rupture of aneurysm of the right sinus of Valsalva into the right ventricular outflow tract: treatment with Amplatzer atrial septal occluder. Circulation 2002; 105: E1-E2.
7. Rao PS, Bromberg BI, Jureidini SB, Fiore AC. Transcatheter occlusion of ruptured sinus of Valsalva aneurysm: innovative use of available technology. Catheter Cardiovasc Interv 2003; 58: 130-4.
8. Fedson S, Jolly N, Lang RM, Hijazi ZM. Percutaneous closure of a ruptured sinus of Valsalva aneurysm using the Amplatzer duct occluder. Catheter Cardiovasc Interv 2003; 58: 406-11.
9. Sakakibara S, Konno S. Congenital aneurysm of the sinus of Valsalva anatomy and classification. Am Heart J 1962; 63: 405-24.
10. Zanchetta M, Rigatelli G, Pedon L, Zennaro M, Maiolino P, Onorato E. Role of intracardiac echocardiography in atrial septal abnormalities. J Interv Cardiol 2003; 16: 63-77.
11. Zanchetta M, Pedon L, Rigatelli G, et al. Intracardiac echocardiography evaluation in secundum atrial septal defect transcatheter closure. Cardiovasc Intervent Radiol 2003; 26: 52-7.
12. Goldberg N, Krasnow N. Sinus of Valsalva aneurysms. Clin Cardiol 1990; 13: 831-6.
13. Edwards E, Burchell HB. The pathological anatomy of deficiencies between the aortic root and the heart, including aortic sinus aneurysms. Thorax 1957; 12: 125-39.
14. Yildirir A, Batur MK, Kabakci G. Ruptured aneurysm of the sinus of Valsalva in association with persistent left superior vena cava: a case report. Angiology 2000; 51: 167-71.
15. Gharzudine WS, Sawaya JI, Kazma HK, Obeid MY. Traumatic pseudoaneurysm of the left sinus of Valsalva: a case report. J Am Soc Echocardiogr 1997; 10: 377-80.
16. van Son JA, 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: II20-II29.
17. Walters MI, Ettles D, Guvendik L, Kaye GC. Intraventricular septal expansion of a sinus of Valsalva aneurysm: a rare cause of complete heart block. Heart 1998; 80: 202-3.
18. Guo DW, Cheng TO, Lin ML, Gu ZQ. Aneurysm of the sinus of Valsalva: a roentgenologic study of 105 Chinese patients. Am Heart J 1987; 114: 1169-77.
19. Xu Q, Peng Z, Rahko PS. Doppler echocardiographic characteristics of sinus of Valsalva aneurysms. Am Heart J 1995; 130: 1265-9.
20. Terdjaman M, Bourdarias JP, Farcot JC. Aneurysms of the sinus of Valsalva. Two-dimensional echocardiographic diagnosis and recognition of rupture into the right heart cavities. J Am Coll Cardiol 1984; 3: 1227-35.
21. Chiang CW, Lin FC, Fang BR. Doppler and two-dimensional echocardiographic features of sinus of Valsalva aneurysm. Am Heart J 1986; 116: 1283-8.
22. Zanchetta M, Onorato E, Rigatelli G, et al. Intracardiac echocardiography-guided transcatheter closure of secundum atrial septal defect. A new efficient device selection method. J Am Coll Cardiol 2003; 42: 1677-82.
23. Holman W. Sinus of Valsalva aneurysms and application of surgical science to their repair. Ann Thorac Surg 1993; 55: 545-51.
24. Naka Y, Kadoba K, Ohtake S, et al. The long-term outcome of a surgical repair of sinus of Valsalva aneurysm. Ann Thorac Surg 2000; 70: 727-9.
25. Au WK, Chiu SW, Mok CK, Lee WT, Cheung D, He GW. Repair of ruptured sinus of Valsalva aneurysm: determinants of long-term survival. Ann Thorac Surg 1998; 66: 1604-10.
26. Abe T, Komatsu S. Surgical repair and long-term results in ruptured sinus of Valsalva aneurysm. Ann Thorac Surg 1988; 46: 520-35.
27. Vural KM, Sener E, Tasdemir O, Bayazit K. Approach to sinus of Valsalva aneurysms: a review of 53 cases. Eur J Cardiothorac Surg 2001; 20: 71-6.