Aneurysms of Sinus of Valsalva: Two-Dimensional Echocardiographic Diagnosis and Recognition of Rupture Into the Right Heart Cavities

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The aim of this study was to evaluate the role of echocardiography in the diagnosis of sinus of Valsalva aneurysms projecting toward the right heart cavities. Three patients who had a ruptured aneurysm of a sinus of Valsalva diagnosed by echocardiography and confirmed by catheterization underwent cardiac surgery. In two patients, the aneurysm originated from the right coronary sinus and had perforated into either the inflow or outflow tract of the right ventricle. In the third patient, the aneurysm, which originated from the noncoronary sinus, ruptured into the right atrium. A fourth patient was also investigated and had an unruptured aneurysm of the right coronary sinus projecting into the right ventricular outflow tract.

M-mode, two-dimensional and contrast echocardiographic studies were performed before cardiac catheterization in all patients and after surgery in three patients. M-mode echocardiography was useful only when the aneurysm had an anterior projection, whether or not the aneurysm was ruptured. Conversely, two-dimensional echocardiography was always able to identify the aneurysmal sac which appeared as an abnormal circular thin-walled structure protruding into the right heart cavities. By using multiple views, it was possible to investigate the whole abnormal structure and locate the sinus from which the aneurysm originated. The use of the echo contrast technique allowed more precise definition of the aneurysmal sac and diagnosis of a left to right shunt by demonstrating a negative contrast image in the right cavities. On the other hand, no negative contrast image was recorded in the patient with an unruptured aneurysm or in the two instances of a successful surgically reconstructed aorta.

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Cardiography was performed in two patients because of a residual murmur.

Case Reports

Between November 1979 and April 1982, four patients were studied. Three had a ruptured sinus of Valsalva aneurysm and underwent surgery; the fourth patient had an unruptured aneurysm and did not undergo surgery.

Case 1: aneurysm of right coronary sinus of Valsalva ruptured into the right ventricular outflow tract. A 21 year old man was hospitalized in 1981 for supraventricular tachycardia and exertional dyspnea. Seven years before, cardiac catheterization performed at another hospital demonstrated a ventricular septal defect with a minor left to right shunt and slight aortic insufficiency. Thereafter, the patient remained asymptomatic.

On admission, a grade 3/6 systolic murmur was heard at the second left intercostal space. On chest X-ray film, the cardiac silhouette was enlarged. The electrocardiogram showed left ventricular hypertrophy. Echocardiography diagnosed an aneurysm of the right sinus of Valsalva that had ruptured into the right ventricular outflow tract. Cardiac catheterization and angiography confirmed the diagnosis.

Perioperative findings revealed an aneurysm of the right aortic sinus with a large perforation (25 × 15 mm) protruding into the right ventricle, a ventricular septal defect and mild aortic insufficiency. Both the ruptured sinus of Valsalva aneurysm and the ventricular defect were closed with a patch; the aortic valve was replaced with a bioprosthesis (Ionescu No. 23). Postoperative echocardiographic study did not demonstrate any residual left to right shunt.

Echocardiography. The preoperative M-mode tracing (Fig. 1) showed an enlarged left ventricle and fluttering of the anterior mitral valve leaflet due to the aortic regurgitation. On the sweep from the left ventricle to the aorta, two linear abnormal echoes corresponding to the walls of the prolapsed aneurysm were recorded in the right ventricular outflow tract and an echo dropout was also noted in the anterior aortic wall. This finding was consistent with the presence of a ruptured sinus of Valsalva aneurysm.

On the parasternal view, two-dimensional echocardiography revealed an abnormal circular structure located just in front of the right sinus of Valsalva, which corresponded to the thin wall of the aneurysmal sac (Fig. 2). Slight tilting of the transducer superiorly and inferiorly allowed delineation of the aneurysm that extended from the level of the anterior portion of the interventricular septum up to the posterior cusp of the pulmonary valve. By rotating the transducer 45° clockwise, the aneurysm was visualized along its long-axis just above the level of the aortic cusps, and an interruption in the echoes was noted at the tip of sac.

After injection of contrast material, a definite negative contrast image arising from the tip of the aneurysm was visualized in the right ventricular outflow tract, mainly during diastole (Fig. 3).

Postoperative echocardiography showed a persistent fluttering of the anterior mitral leaflet, but the aneurysmal sac was no longer visible (Fig. 4). Contrast studies demonstrated the lack of residual left to right shunt.

Case 2: aneurysm of the right coronary sinus of Valsalva ruptured into the right ventricular inflow tract. A 59 year old man was hospitalized for investigation of severe congestive heart failure evolving for 4 months. Physical examination revealed a grade 3/6 diastolic murmur and a grade 2/6 systolic murmur heard over the entire precordium.

Figure 1. Patient 1. Preoperative M-mode sweep from the left ventricle (LV) (left) to the aortic (Ao.) root (right). The right ventricle (RV) is normal but the left ventricle is enlarged. The anterior mitral valve leaflet (AMV) exhibits diastolic fluttering (f). There is a herniation (arrow 1) of the right coronary cusp into the right ventricular outflow tract, along with an abnormal two-layered echo (arrows at 2) representing the aneurysmal sac.
Figure 2. Patient 1. **Upper panels,** Two-dimensional parasternal cross-sectional short-axis views of the base of the heart in systole (S) and in diastole (D). Just above the aorta (Ao), in the vicinity of the right coronary sinus, there is a thin, round, additional echo (arrow) that points to the aneurysmal sac protruding toward the right ventricular outflow tract (RVOT). In diastole, the sac is larger (arrow). **Lower panels,** By slightly rotating the transducer in a clockwise manner, the aneurysm of sinus of Valsalva (arrow) is well delineated in systole (S), as well as in diastole (D), just above the level of the aortic cusps (c). LA = left atrium; LV = left ventricle; RA = right atrium.

Blood pressure was 170/80 mm Hg. The electrocardiogram showed right bundle branch block. The echocardiographic study revealed an aneurysm of the right sinus of Valsalva ruptured into the right ventricle and an associated aneurysm of the atrial septum.

**Cardiac catheterization and surgery confirmed the diagnosis.** The aneurysm had perforated into the right ventricle just below the septal leaflet of the tricuspid valve. In addition, a patent foramen ovale was found. Postoperatively, a continuous murmur was noted and congestive heart failure recurred. Echocardiography demonstrated a dehiscence of the pericardial patch and a residual left to right shunt. Because the hemodynamic condition was deteriorating, the patient underwent reoperation and a 4 mm disruption of the patch was repaired.

**Echocardiography.** The preoperative M-mode studies revealed moderate left ventricular enlargement. No signs of aortic regurgitation and no premature pulmonary valve opening were noted. Two-dimensional cross-sectional short-axis views were performed at three levels. At the mitral valve level (Fig. 5A), a thin-walled circular structure was tangentially attached to the right side of the interventricular septum. At the level of the base of the heart (Fig. 5B), there was an obvious systolic-diastolic defect of the aortic ring just in front of the right anterior aortic cusp. In a slightly superior plane (Fig. 5C), the aortic root anatomy appeared normal in systole and slightly enlarged in diastole. Because of the associated shunt at the atrial level, satisfactory contrast studies were not obtained in this patient, but this well delineated wind-scock shaped image was highly suggestive of an aneurysm of the right coronary sinus of Valsalva protruding into the right anulus (Fig. 5B).

**Postoperative two-dimensional echocardiography** clearly identified the pericardial patch closing the aortic defect. Although the aortic ring appeared normal (Fig. 6, left), a diastolic negative contrast image was demonstrated in front of the patch (Fig. 6, right) indicating its partial dehiscence.

**Case 3: aneurysm of the noncoronary sinus of Valsalva ruptured into the right atrium.** A 71 year old man was referred to our institution because of bacterial endocarditis. A grade 4/6 systolic-diastolic murmur was heard
Echocardiography. The preoperative M-mode echocardiogram showed a normal apex to base sweep, right ventricular enlargement and fluttering of the anterior mitral valve leaflet. By using a parasternal approach, two-dimensional echocardiography revealed an abnormal aortic root with a dilation of the posterior sinus of Valsalva. There was a disruption in the continuity of the aortic ring facing the right atrium in the vicinity of the tricuspid valve (Fig. 7A). The same image was recorded in a slightly modified apical four chamber view visualizing the posterior sinus of Valsalva (Fig. 7B). Contrast studies confirmed the perforation by showing an obvious negative contrast image, mainly during diastole, due to the unopacified blood shunting from the noncoronary sinus into the right atrium (Fig. 7C).

Case 4: unruptured aneurysm of the right coronary sinus of Valsalva protruding into the right ventricular outflow tract. A 49 year old man was hospitalized for exertional atypical chest pain and palpitation evolving for 10 months. On physical examination, a grade 2/6 early systolic murmur was heard at the fifth left intercostal space. The electrocardiogram showed large inverted asymmetric T waves in leads V4 to V6. Echocardiography demonstrated an aneurysm of the right sinus of Valsalva, and contrast injection excluded a left to right shunt. Selective aortography imaged an unruptured saccular dilation of the right sinus of Valsalva. Coronary angiography was normal and an ergonovine test was negative for coronary spasm. Left ventriculography was normal.

Echocardiography. On M-mode echocardiography, the apex to base sweep exhibited an enlargement of the aorta over the entire precordium. Severe congestive heart failure was present. The electrocardiogram showed complete atrioventricular block with a ventricular rate at 40 beats/min that required temporary pacing. Echocardiography revealed the presence of an aortic valve vegetation and an aneurysm of the noncoronary sinus that had ruptured into the right atrium.

Cardiac catheterization and aortography confirmed the diagnosis. After a 12 day period of massive antibiotic therapy, the patient underwent surgery. The aneurysm originated from the noncoronary sinus and was protruding into the right atrium close to the tricuspid anulus. The perforation was ovular in shape, 12 mm long and 8 mm wide. The aneurysm was repaired with a Dacron patch and the aortic valve was replaced. The patient died 15 days later of recurrent septic shock.

Figure 3. Patient 1. Two-dimensional echocardiographic cross-sectional short-axis views of the base of the heart (left) showing in diastole the aneurysm (An) of sinus of Valsalva described in Figure 2. Right panel, Contrast study reveals a permanent diastolic negative contrast effect due to the left to right shunt (arrow). LA = left atrium; RA = right atrium.

Figure 4. Patient 1. Postoperative echocardiograms showing no abnormal echo on the cross-sectional views of the base of the heart (A) in the right ventricular outflow tract (RVOT). In the aortic root, the struts of the prosthetic Ionescu valve (Iv) are well visualized. On a slightly inferior plane (B) at the level of the mitral valve (MV), the aneurysmal sac is no longer visible. In the right panel (C), contrast echocardiographic study excludes any residual left to right shunt. LA = left atrium; RA = right atrium.
with an abnormal motion pattern of the unusually thin anterior aortic wall (Fig. 8). In addition, the right coronary cusp had an early systolic closure and maintained a distance from the anterior wall. An abnormal mobile echo was seen in the right ventricular outflow tract anterior to the anterior

Figure 5. Patient 2. Cross-sectional short-axis views at three different levels. A, mitral valve (MV); B, sinuses of Valsalva and C, just above the latter. In A, the aneurysm (An) is clearly seen during systole (S) and diastole (D) protruding into the right ventricular (RV) inflow tract. In C, the aortic root anatomy appears normal during systole and slightly enlarged during diastole. LA = left atrium; RA = right atrium.

Figure 6. Patient 2. Postoperative cross-sectional short-axis views obtained at the same level as in Figure 5B. Although the aortic root appears well reconstructed (left panel), the aneurysmal sac free walls are still visible. A negative contrast (nc) effect due to residual left to right shunt is demonstrated just in front of the patch. LA = left atrium; RA = right atrium; RV = right ventricle.
Figure 7. Patient 3. A, Parasternal cross-sectional short-axis view of the base of the heart demonstrating a disruption in the continuity of the aortic (AO) ring facing the right atrium (RA) in the vicinity of the tricuspid valve (t) which represents the ruptured aneurysm (An) of the noncoronary sinus of Valsalva. B, Slightly modified apical four chamber view imaging the noncoronary sinus of Valsalva and showing the ruptured aneurysm (arrow). C, Negative contrast (nc) obtained in the same view as in B, representing the unopacified blood shunting into the right atrium just above the tricuspid valve (arrow). LA = left atrium; LV = left ventricle; RV = right ventricle.

aortic wall and was still present on the sweep down to the basal portion of the interventricular septum.

Parasternal short-axis cross-sectional views (Fig. 9) were obtained at three levels between the mitral valve and the aortic valve. They showed normal cardiac anatomy at the level of the free edges of the mitral valve (Fig. 9A). In a slightly higher plane, an abnormal thin-walled circular structure protruding into the right ventricle (Fig. 9B) was clearly seen anterior to the right coronary sinus. At the level of the aortic ring, dilation of the right coronary sinus was well defined during diastole (Fig. 9C). Contrast studies allowed precise delineation of the shape of the aneurysm and exclusion of a left to right shunt (Fig. 9, right panels D to F).

Discussion

Classification of sinus of Valsalva aneurysms. The clinical symptoms of a ruptured sinus of Valsalva aneurysm depend on both its location and the magnitude of the shunt. In the early 1960s, an anatomic classification of the various congenital aneurysms of the sinus of Valsalva was attempted by Sakakibara and Konno (5). They standardized four types according to the location of the aneurysm and the site of the rupture. Another classification based on the direction of the extension toward the right heart cavities was proposed by Acar and Cartier (6). Two types can be individualized: 1) the aneurysm projects posteriorly and ruptures into either the right atrium or the right ventricular inflow tract in the vicinity of the tricuspid valve, and it originates from the right coronary or the noncoronary sinus; and 2) the aneurysm projects anteriorly and ruptures into the right ventricular outflow tract just beneath the pulmonary cusp, and it originates exclusively from the right coronary sinus. An interventricular septal defect is associated in approximately 50% of the latter type of aneurysm. By virtue of the capability of echocardiography to study the spatial relation of the right heart cavities to the three sinuses of Valsalva, we used echocardiography coupled with the contrast technique to study the location, extension and possible rupture in four consecutive cases of sinus of Valsalva aneurysm.

M-mode echocardiography. Numerous M-mode echocardiographic abnormalities have been reported in patients with a sinus of Valsalva aneurysm. Some of these findings are not specific, such as the early systolic closure of the aortic cusps (7) or the presence of abnormal echoes in the left or right atrium (8-10) . Other echocardiographic findings are more meaningful: 1) Weyman et al. (10) reported the highly suggestive association of an echo-producing mass in the right atrium and an early diastolic pulmonary valve opening, the latter being usually observed in adiastole or valvular pulmonary stenosis (10); 2) a high frequency fluttering of the anterior tricuspid leaflet both in early diastole and systole due to the jet stream occurring throughout the cardiac cycle (11); and 3) a diastolic murmur without fluttering of the anterior mitral leaflet.

When recorded on a sweep from the aorta to the left ventricle, the following are considered to be highly suggestive of a right coronary sinus aneurysm: 1) a large echo-free area anterior to the aortic valve representing the aneurysmal dilation (11-13); 2) abnormal echoes extending from the aortic root into the left ventricular outflow tract and presumed to represent the prolapsed aneurysmal sac (14); and 3) pattern of the interventricular septum that appears separated into two distinct parallel echoes approximately at the level of the anterior mitral leaflet (15). In our patients, only the extension of the right coronary sinus aneurysm was
Figure 8. Patient 4. M-mode echocardiographic base to apex sweep of a proven unruptured aneurysm of the right coronary sinus of Valsalva protruding into the right ventricular outflow tract (RVOT). The right coronary cusp of the aorta (AO) has an early systolic closure (arrow 1) and stays away from the anterior aortic wall. An abnormal ill-defined echo (arrows at 2) is present in the right ventricular (RV) outflow tract. Note the similarity of this abnormal echo with the one depicted in Figure 1, also showing an aneurysm of the right coronary sinus but one with rupture into the right ventricular outflow tract. LA = left atrium; LV = left ventricle.

suggested by M-mode tracings and this finding is in accord with previous reports (11,13,16). Because of the anterior and superior projection of the aneurysm, the M-mode ultrasonic beam can detect only those aneurysms that project toward the right ventricular outflow tract. Indeed, M-mode echocardiography failed to diagnose the aneurysm in our Cases 2 and 3 because the aneurysm had a more lateral extension toward the right ventricular inflow tract or the right atrium.

Two-dimensional echocardiography. Spatial orientation capabilities of two-dimensional echocardiography enable identification of different types of sinus of Valsalva aneurysms. Although a complete two-dimensional echocardiographic examination is recommended, cross-sectional short-axis views at the level of the base of the heart easily portray the three sinuses of Valsalva, especially during diastole when each aortic cusp can be identified (17). By slightly tilting the transducer to record cross sections in different planes from the origin of the aneurysm to the distal portion of the sac, its extension and the site of the rupture can be fully investigated. Direct visualization of the aneurysm of sinus of Valsalva rupturing into either the right ventricular outflow tract (11–13,16) or the right atrium (18) has been reported using the parasternal views.

In our four patients, using the parasternal views, it was possible to precisely identify the aortic sinus from which the aneurysm originated. Two patients had an aneurysm of the right coronary sinus that had ruptured into the right ventricle beneath the pulmonary cusp (Patient 1) or near the tricuspid valve (Patient 2). Patient 3 demonstrated a posterior sinus of Valsalva aneurysm rupturing into the right atrium. The remaining patient (Patient 4) had an unruptured aneurysm of the right coronary sinus extending toward the right ventricular outflow tract.

Although most reports claim that two-dimensional echocardiographic findings were confirmed in patients by surgery, studies on sensitivity and specificity of two-dimensional echocardiography are still lacking. In the presence of an aneurysm of the sinus of Valsalva diagnosed by two-dimensional echocardiography, accurate identification and location of the rupture requires the demonstration of a discontinuity in the echoes arising from the tip of the aneurysm, as was the case in three of our four patients. The possibility of false positive (echo dropouts) or false negative (due to the limited lateral resolution) findings must be kept in mind. Therefore, even when the rupture is large and directly visualized, the echo contrast technique is essential to diagnose the left to right shunt (16,18).

Contrast echocardiography. The echo contrast technique may be very helpful in analyzing unusual echo structures, the precise identification of which is difficult. The presence of microcavitations in the right heart allows differentiation of structures belonging to the right or the left heart cavities, respectively. Thus, the contours of the aneu-
rysm of the sinus of Valsalva may be clearly encircled by the microbubbles (Fig. 9). In some cases, this technique may help ascertain whether an unidentified echo structure corresponds to an aneurysm of the membranous septum or an aneurysm of the sinus of Valsalva. Furthermore, differentiation between echo dropouts and actual structural cardiac discontinuity, on one hand, and identification of the left to right shunt, on the other hand, are fundamental. Schatz et al. (16) and Nakamura et al. (18) described the use of two-dimensional contrast echocardiography to detect rupture of the sinus of Valsalva aneurysm into the right ventricular outflow tract or into the right atrium by recording the negative jet during diastole in these chambers.

In three of our four patients, two-dimensional echocardiography visualized a permanent discontinuity of the aneu-

Figure 9. Patient 4. Standard cross-sectional short-axis views (left panels) and contrast studies (right panels) obtained at the level of the free edges of the mitral valve (A), in a slightly upper plane (B) and at the level of the aortic ring (C). Left panels, In A, both ventricles appear normal; in B, the aneurysmal sac (An) is well delineated and protrudes into the right ventricular outflow tract (RVOT); in C, moderate dilation of the right coronary sinus is visualized. Right panels, Contrast studies performed in the same planes delineated the shape and the small size of the aneurysm (D and E) and excluded any left to right shunt originating from the aneurysm (E and F). LA = left atrium; LV = left ventricle; RA = right atrium; RV = right ventricle.
three patients: toward the right ventricular outflow tract in Patient 1 and the right atrium near the tricuspid valve in Patient 3. In contrast, in Patient 4 who was suffering from an unruptured aneurysm of the right coronary sinus protruding into the right ventricular outflow tract, contrast studies excluded an associated perforation. Lastly, in Patient 2, preoperative contrast echocardiography failed to demonstrate a left to right shunt at the ventricular level, probably because an associated shunt at the atrial level impeded a satisfactory opacification of the right ventricle. However, in this patient, postoperative contrast studies identified residual shunting due to partial dehiscence of the pericardial patch closing the aortic defect. This complication could not have been detected by standard two-dimensional echocardiography.

Potential limitations can affect the identification of the left to right shunt. The jet of blood across the ruptured aneurysm of sinus of Valsalva may be directed outside of the echocardiographic plane being viewed; furthermore the negative contrast image, usually evidenced during diastole, may be so transient that it is missed, especially in the case of tachycardia. Lastly, the presence of an upstream shunt may impede adequate opacification of the right heart cavities. Therefore, an examination using numerous echo planes, multiple contrast injections and a frame by frame analysis is needed.

Conclusion. The contribution of M-mode echocardiography to the diagnosis of aneurysms of the sinus of Valsalva is limited to those with anterior extension. Aneurysms that project posteriorly are not visualized by M-mode investigations. Wide angle two-dimensional echocardiographic techniques proved to be of great benefit in delineating the size and shape of the aneurysmal sac, demonstrating the site of the rupture, identifying the left to right shunt and differentiating the Valsalva aneurysm from the other causes of continuous murmurs. The ability of two-dimensional echocardiography associated with contrast studies to accurately define the anatomic lesions before cardiac catheterization and surgery suggests that this technique should be recommended in the management of patients suspected of sinus of Valsalva aneurysm. In addition, two-dimensional echocardiography allows postoperative control of these patients.

We are grateful for the expert secretarial assistance of Catherine Eichner, Anges Autef-Gauthier and Myriam Briez.

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