Introduction and objectives. Ruptured sinus of Valsalva aneurysm to right cardiac chambers is an uncommon lesion in Western countries. The prognosis is usually serious unless the condition is promptly treated surgically. For this reason an accurate anatomical and functional evaluation is necessary. The main purpose of this report is to compare the usefulness of multiplane transesophageal echocardiography with transthoracic echocardiography and angiocardiography in the preoperative evaluation of ruptured sinus of Valsalva aneurysm to right chambers.

Patients and method. Since January 1990, 9 patients (mean age 36.3 ± 18 years, 6 males) with ruptured sinus of Valsalva aneurysm to right chambers were studied. The pathogenesis was congenital aneurysm in 6 patients, aortic prosthesis endocarditis in one and two cases of iatrogenia: during a percutaneous mitral valvuloplasty and after cardiac surgery. Transthoracic echocardiography was performed in all cases, transesophageal echocardiography in 7 and angiocardiography in 8. Two patients died before surgery, and 7 were successfully operated on.

Results. Transesophageal echocardiography was more useful when compared to transthoracic echocardiography and angiocardiography in detecting: a) the fistula; b) the sinus involved; c) the right chamber affected; d) congenital aneurysms morphology and size; e) aneurysm prolapse through a ventricular septal defect, and f) the identification of other cardiac congenital or acquired anomalies.

Conclusions. Multiplane TEE is the most accurate tool in the preoperative evaluation of ruptured sinus of Valsalva aneurysm to right chambers.

Key words: Congenital heart diseases. Fistula. Echocardiography. Surgery.

INTRODUCTION

Sinus of Valsalva fistulas to right chambers (SVFRC) are an infrequent pathology in surgical series in Western countries,1-3 their prevalence is much...
higher in Asian populations. Once it has developed, the fistula originates a rapidly progressive left-to-right shunt that generally is manifested by congestive heart failure refractory to medical treatment. The ruptured aneurysm can be repaired surgically with excellent immediate and long-term results, so surgery should be performed without delay to prevent more morbidity and mortality. The diagnosis may be suspected on the basis of clinical manifestations and confirmed by cardiac catheterization and aortography, but in many cases AC does not clearly define the underlying defect or accompanying lesions. Recently it has been reported that TEE can be very useful for the preoperative assessment of patients with this disease. The purpose of this study is to compare the effectiveness of TEE with TTE and AC in the diagnosis and preoperative assessment of patients with SVFRC.

PATIENTS AND METHOD

Study population

From January 1990 to October 2000, 9 patients with SVFRC were documented in our service. The diagnosis was confirmed in the surgical intervention in 7 patients, but 2 patients did not undergo surgery and the diagnosis was based only on echocardiographic and angiographic findings. The clinical and surgical protocols, echocardiographic and angiohemodynamic findings, and long-term evolution of all patients were reviewed.

Echocardiographic assessment

TTE was performed in all patients and multiplane TEE was carried out in. The studies were made with Sonos 2000, 2500 or 5500 equipment (Agilent Technologies, Andover, MA). TTE was performed with a 2.5-MHz probe or a multifrequency probe, with or without second harmonic imaging. The standard planes were studied from the parasternal, apical, and subcostal positions with bidimensional and Doppler color imaging. The size and morphology of the aortic root, degree of insufficiency of the aortic valve, and existence of a shunt between the aortic root and right chambers were assessed. The presence of accompanying lesions, especially malformations of the aortic valve or sinuses of Valsalva, endocardial vegetations, and defects in the interventricular septum, was evaluated. The size and function of the cardiac chambers were measured in all patients using the M-mode technique guided by the bidimensional image, and the systolic pulmonary artery pressure and pulmonary-to-systemic blood flow ratio were estimated using previously established methods.

Multiplane TEE was carried out with a multiplane probe and multifrequency transducer of 5/3.5 MHz with pulsed Doppler, continuous, and color capabilities. The probe was placed in the esophagus and the cross-sectional plane of the aortic root was analyzed using a transducer rotation of 30° to 45°, and the longitudinal plane with a transducer rotation of 120° to 150°. In the cross-sectional plane the morphology of the three sinuses of Valsalva and their relation with neighboring structures was analyzed, especially the right ventricular outflow tract, right atrium, tricuspid valve, and interatrial septum. In the longitudinal plane, the anatomy of the right coronary and non-coronary sinuses of Valsalva, their relation with the right ventricular outflow tract and left atrium, and the degree of aortic valve insufficiency were analyzed. The transducers were rotated continuously between the two planes described, varying the anteflexion and probe depth to better assess the anatomy of the aortic root and related structures. In addition, a transgastric examination was made to localize the outflow tracts of both ventricles in a disposition longitudinal to the ultrasonic beam in order to assess the infundibular septum, degree of aortic valve insufficiency, and pressure gradient in the right ventricular outflow tract.

RESULTS

Pathogenesis and clinical manifestations

The mean age of patients was 36.3 ± 18 years (range, 12-58 years). There were 6 men and 3 women. The fistula originated in the right sinus of Valsalva in 6 cases and in the non-coronary sinus in 3. In 4 patients the fistula communicated the aortic root with the right ventricular outflow tract, and in the other 5 patients it opened onto the right atrium. All the fistulas that opened onto the right ventricular outflow tract originated in the right coronary sinus of Valsalva, but of the 5 fistulas communicating with the right atrium, 2 originated in

ABBREVIATIONS

SVFRC: sinus of Valsalva fistula to right chambers.
TEE: transesophageal echocardiography
TTE: transthoracic echocardiography.
AC: angiography.
VSD: ventricular septal defect
the right sinus of Valsalva and 3 in the non-coronary sinus. The cause of the fistula was rupture of a congenital aneurysm in 6 cases, aortic endocarditis in 1, and iatrogenesis in 2.

Two patients had a VSD associated with a subarterial fistula. One patient had a bicuspid aortic valve and previously corrected aortic coarctation. The other patient had severe subvalvular pulmonic stenosis due to a fibromuscular band. In one patient the fistula was produced by prosthetic aortic valve endocarditis. Of the patients with an iatrogenic fistula, one originated during attempted percutaneous mitral valvuloplasty for severe mitral stenosis, and the other was the consequence of aortic valve replacement. In the latter case, extensive dissection of the interatrial septum occurred in the postoperative phase, forming a false chamber that perforated the right atrium.

In 8 patients, the fistula produced a large left-to-right shunt with a Qp/Qs ratio > 2. Only the patient with an iatrogenic fistula after mitral valvuloplasty had a small shunt, with Qp/Qs < 1.5. The 8 patients with a large left-to-right shunt presented symptoms of congestive heart failure as the initial manifestation, with symptoms of right heart failure predominating over those of left heart failure. The cause of the fistulas, main lesions associated, and clinical pictures are summarized in table 1. Two patients died before surgery: one patient, who refused surgery, died of congestive heart failure a few months after diagnosis; the other patient, who had prosthetic valve endocarditis, died of sepsis, cerebral embolism, and multiorgan failure. The remaining 7 patients were treated surgically as priority patients. No perioperative complications occurred. All the patients who underwent surgery survived the intervention and are alive after a mean follow-up time of 48 ± 34 months.

Echocardiographic findings

TEE detected a shunt between the aortic root and right chambers in 7 patients (78%) and was valuable in determining the size and function of the cardiac cavities, degree of aortic insufficiency, systolic pulmonary artery pressure, and pulmonary-to-systemic flow ratio. However, it was less effective in identifying the beginning and end of the fistula, or in diagnosing congenital aneurysm and its morphology. Of the 2 cases of subarterial VSD, TTE identified only one (Table 2).

### TABLE 1. Etiopathogenesis and clinical course in 9 patients with sinus of Valsalva fistula to right chambers

<table>
<thead>
<tr>
<th>Case no.</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Sinus of Valsalva</th>
<th>Drainage chamber</th>
<th>Etiology</th>
<th>Associated lesions</th>
<th>Clinical manifestations</th>
<th>Surgery</th>
<th>Duration of follow-up (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>21</td>
<td>F</td>
<td>RC</td>
<td>RV</td>
<td>Congenital aneurysm</td>
<td>VSD</td>
<td>CHF</td>
<td>Yes</td>
<td>92</td>
</tr>
<tr>
<td>2</td>
<td>58</td>
<td>F</td>
<td>NC</td>
<td>RA</td>
<td>Mitral valvuloplasty</td>
<td>Mitral stenosis</td>
<td>Asymptomatic</td>
<td>Yes</td>
<td>60</td>
</tr>
<tr>
<td>3</td>
<td>35</td>
<td>F</td>
<td>RC</td>
<td>RV</td>
<td>Congenital aneurysm</td>
<td>DCRV</td>
<td>CHF</td>
<td>Yes</td>
<td>48</td>
</tr>
<tr>
<td>4</td>
<td>13</td>
<td>M</td>
<td>RC</td>
<td>RA</td>
<td>Congenital aneurysm</td>
<td>Bicuspid aorta</td>
<td>CHF</td>
<td>Yes</td>
<td>12</td>
</tr>
<tr>
<td>5</td>
<td>12</td>
<td>M</td>
<td>RC</td>
<td>RV</td>
<td>Congenital aneurysm</td>
<td>VSD</td>
<td>CHF</td>
<td>Yes</td>
<td>36</td>
</tr>
<tr>
<td>6</td>
<td>38</td>
<td>M</td>
<td>NC</td>
<td>RA</td>
<td>Congenital aneurysm</td>
<td>No</td>
<td>CHF</td>
<td>No</td>
<td>Death</td>
</tr>
<tr>
<td>7</td>
<td>54</td>
<td>M</td>
<td>RC</td>
<td>RA</td>
<td>Congenital aneurysm</td>
<td>No</td>
<td>CHF</td>
<td>Yes</td>
<td>3</td>
</tr>
<tr>
<td>8</td>
<td>57</td>
<td>M</td>
<td>RC</td>
<td>RV</td>
<td>Prosthetic valve endocarditis</td>
<td>Aortic prosthesis</td>
<td>CHF</td>
<td>No</td>
<td>Death</td>
</tr>
<tr>
<td>9</td>
<td>39</td>
<td>M</td>
<td>NC</td>
<td>RA</td>
<td>Interal atrial septal dissection</td>
<td>Aortic prosthesis</td>
<td>CHF</td>
<td>Yes</td>
<td>84</td>
</tr>
</tbody>
</table>

CHF indicates congestive heart failure; DCRV, double-chambered right ventricle; NC, non-coronary; RA, right atrium; RC, right coronary; RV, right ventricle; VSD, ventricular septal defect.

### TABLE 2. Comparison of transthoracic echocardiography (TTE), transesophageal echocardiography (TEE), and angiocardiography (AC) in the diagnosis and preoperative assessment of the sinus of Valsalva fistulas perforated in right cardiac chambers

<table>
<thead>
<tr>
<th>TEE</th>
<th>TTE</th>
<th>AC</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diagnosis of the fistula</td>
<td>7/7 (100%)</td>
<td>7/9 (78%)</td>
</tr>
<tr>
<td>Location of the sinus of Valsalva involved</td>
<td>7/7 (100%)</td>
<td>7/9 (78%)</td>
</tr>
<tr>
<td>Location of the drainage chamber</td>
<td>7/7 (100%)</td>
<td>6/9 (67%)</td>
</tr>
<tr>
<td>Definition of a congenital aneurysm</td>
<td>4/4 (100%)</td>
<td>3/6 (50%)</td>
</tr>
<tr>
<td>Morphology and size of the aneurysm</td>
<td>4/4 (100%)</td>
<td>1/6 (17%)</td>
</tr>
<tr>
<td>Diagnosis of associated anomalies</td>
<td>6/6 (100%)</td>
<td>3/7 (43%)</td>
</tr>
</tbody>
</table>

AC: angiocardiography; TEE: transesophageal echocardiography; TTE: transthoracic echocardiography. The numbers indicate positive findings/patients studied.
TEE (Table 2) identified the fistula, sinus of Valsalva involved, and drainage orifice of the fistula in the right atrium or right ventricle in all the cases (Figure 1). The most useful view was the cross-sectional plane of the aortic root obtained with a rotation of 30° to 60°. In this plane, the morphology and number of aortic leaflets and location of the fistula could always be visualized (Figure 2). The morphology and size of the congenital aneurysm were clearly determined in every case. In the only patient with VSD in which TEE was performed, the prolapse of the aneurysm through the septal defect was visible (Figure 3). The aneurysm that prolapsed through the VSD and perforated the right ventricular outflow tract had a special mobility. In diastole it was spherical because the pressure in the aortic root was greater, but in systole it collapsed under the pressure of flow from the left ventricle through the VSD (Figure 3). In contrast, aneurysms not associated with a VSD had a spherical morphology in both systole and diastole because the pressure is higher in the aortic root than in the drainage chamber throughout the cardiac cycle (Figure 2). Finally, TEE made it possible to delimit other coexisting anomalies, such as subvalvular pulmonic stenosis or patent foramen ovale and, in the case of endocarditis, the perivalvular extension of the infection. The findings of TEE were confirmed in all the patients who underwent surgery.

In comparison with AC, TEE also was more effective in: a) diagnosing the fistula and differentiating it from a sinus of Valsalva prolapse through a VSD (syndrome of Laubry); b) identifying the sinus of Valsalva affected and the cardiac drainage chamber; c) determining the cause of the fistula and the underlying mechanism; d) visualizing the morphology of congenital aneurysms, and e) defining the associated congenital or acquired cardiac anomalies (Table 2).

DISCUSSION

This study describes the etiopathogenesis, clinical
course, and diagnosis of 9 patients with SVFRC followed-up in our center in the last 10 years, with special emphasis on the usefulness of multiplane TEE in the preoperative assessment of this entity.

**Etiopathogenesis**

Sinus of Valsalva fistula to right chambers is an uncommon entity in the Western world.\(^1\)\(^-\)\(^5\) We found 9 cases in almost 50,000 patients studied echocardiographically in the same period, a prevalence of less than 0.02%. The prevalence is much higher in Asian countries, ranging from 0.96 to 1.5% of cardiac surgery procedures with extracorporeal circulation\(^4\) and reaching 0.2% of all echocardiographic studies.\(^8\)

The most frequent cause of SVFRC is perforation of a congenital sinus of Valsalva aneurysm, but other causes have been described, including infections (endocarditis, luetic aortitis) and traumatic or iatrogenic dissection.\(^1\)\(^-\)\(^6\),\(^11\),\(^12\) In our series, the underlying cause was congenital aneurysm of a sinus of Valsalva in 6 patients (67%), infective (prosthetic valve endocarditis) in 1 patient, and iatrogenic in 2: percutaneous mitral valvuloplasty and dissection of the interatrial septum after surgery for aortic valve replacement. To our knowledge, these two iatrogenic causes of sinus of Valsalva fistula to right chambers have not yet been described.

Most patients with SVFRC have a rapidly progressive, voluminous left-to-right shunt that is poorly tolerated. Eight of our patients had a large shunt with Qp/Qs > 2. The clinical presentation was similar in all the patients, consisting of symptoms and signs of predominantly right heart failure. Given the gravity of the clinical condition, surgical intervention should not be delayed. Before surgery, diagnostic techniques should be used to confirm the clinical suspicion, differentiate it from other causes of continuous murmur,\(^13\),\(^14\) and obtain as much exact information as possible about the origin and destination of the fistula, underlying cause, and effect on function ventricular.

**Associated lesions**

In patients with congenital aneurysm, associated lesions are not uncommon. Two of our patients had
a subarterial VSD, 1 had aortic coarctation and bicuspid aortic valve, and 1 had subinfundibular pulmonic stenosis (double-chambered right ventricle). The frequent association of these congenital anomalies, especially VSD, has been reported in other series.\(^{3,4,6-8}\) These congenital anomalies make the preoperative diagnostic assessment more difficult. Another factor of diagnostic confusion is the presence of aortic insufficiency. Two patients in our series had severe aortic insufficiency dependent on the aneurysm. Aortic valve insufficiency is common in patients with sinus of Valsalva aneurysm that has ruptured into the right chambers, regardless of the cause. In at least 10% to 20% of cases, aortic insufficiency requires prosthetic valve replacement or aortic valvuloplasty at the time of surgery.\(^{3,6,7}\)

### Cardiac catheterization

The use of different diagnostic methods in SVFRC has changed in recent decades. Initially, AC was the fundamental diagnostic method.\(^{15}\) In 1967 De Bakey\(^{16}\) described a series of 35 cases of fistulated sinus of Valsalva aneurysms diagnosed by aortography. In 1975, Meyer et al\(^{1}\) published a series of 45 patients who underwent surgery after catheterization and AC, which was inconclusive in 15 cases. AC had many limitations in our series. It did not detect the fistula in 2 cases and was not very useful in precisely identifying the drainage chamber of the fistula into the right heart and the sinus of Valsalva affected. It also failed to delimit the associated lesions in 2 patients. However, cardiac catheterization and AC were very useful in assessing aortic insufficiency and determining pulmonary pressure and resistance and the degree of left-to-right shunt.

### Transthoracic echocardiography

In 1974, Rothbaum et al\(^{17}\) reported echocardiographic findings of ruptured sinus of Valsalva aneurysms. Since then,\(^{18-27}\) numerous publications have appeared, demonstrating the usefulness of bidimensional and Doppler echocardiography in the assessment of these fistulas. In our series, conventional echocardiography with Doppler color was performed in all patients and initially allowed the identification of a shunt between the aortic root and right chambers in 78% of the cases. TTE also was very useful in assessing the hemodynamic repercussions (Qp/Qs, pulmonary artery pressure) and degree of associated aortic insufficiency. However, it was insufficient in determining the size and morphology of the congenital aneurysms, exactly delimiting the beginning and end of the fistula, and diagnosing other associated congenital or acquired lesions. Chiang et al\(^{27}\) demonstrated that bidimensional TTE failed to detect up to 42% of congenital sinus of Valsalva aneurysms, although this deficit could be reduced to 25% with contrast injection. Doppler color increases the sensitivity of transthoracic echocardiography, but it may fail to detect smaller or unruptured aneurysms. The greatest limitation of this technique is difficulty in identifying VSD because it masks flows.\(^{8}\) Nonetheless, several publications\(^{3,28}\) have demonstrated that surgical repair can be carried out in selected patients using bidimensional and Doppler echocardiography studies, without need for previous catheterization.

### Multiplane transesophageal echocardiography

The usefulness of TEE in the preoperative assessment of SVFRC has been demonstrated in publications of isolated cases.\(^{12,23,29-37}\) Its usefulness has been compared to conventional echocardiography and angioangiography in only one Asian series of 23 patients in which multiplane TEE was performed in 10 cases.\(^{38}\) In our series preoperative TEE was performed in 7 cases. This technique proved highly sensitive in the identification of the fistula, sinus of Valsalva involved, and cardiac drainage chamber. The size and morphology of the congenital aneurysm could be evaluated in all the cases. In addition, TEE easily differentiated isolated perforation of a congenital sinus of Valsalva aneurysm and perforation of an aneurysm prolapsed through a subarterial VSD. The aneurysm prolapsed through the VSD had a very different morphology in systole and diastole. During diastole the aneurysm was distended and spherical due to the higher pressure in the aortic root, but during systole the aneurysm collapsed due to the high-pressure shunt from the left to the right ventricle (Figure 3). This dynamic behavior of sinus of Valsalva aneurysms perforated through a VSD has not been described previously. TEE was also more useful in assessing other associated congenital lesions, but it was not superior to TTE or aortography in assessing the degree of aortic insufficiency or left-to-right shunt.\(^{38}\)

### CONCLUSIONS

SVFRC are an entity with a very low prevalence, frequently congenital, and in most cases require early surgical treatment. This diagnosis should be suspected in patients with a continuous cardiac mur-
murmur, particularly if symptoms of congestive heart failure of rapid onset are present. Bidimensional echocardiography with Doppler color in most cases can confirm the diagnosis and provide a hemodynamic assessment of the degree of left-to-right shunt and/or aortic insufficiency. Multiplane TEE is the most useful diagnostic method for the assessment of the etiopathogenesis and anatomy of these fistulas, facilitating the election of the optimal surgical approach and treatment. Although cardiac catheterization has been shown to be less effective in diagnosing SVFRC, it continues to be useful in selected patients. However, it should not be considered the first-choice diagnostic method, but instead a complementary technique.

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