

## Case reports

## Ruptured Aneurysm of the Right Sinus of Valsalva Into the Right Ventricle

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**Aneurysm of sinus of Valsalva, rupture.**

Aneurysms of sinus of Valsalva, are rare cardiac abnormalities with congenital origin in most of the cases. If they are located in the right coronary sinus, they usually rupture into right heart chambers and frequently a ventricular septal defect coexists with this condition. We present a case of a 42 year old man with ruptured aneurysm of the right sinus of Valsalva into the right ventricle. Transesophageal echocardiographic examination in addition to transthoracic echocardiography provides more powerful information about the aneurysms and coexistent cardiac malformations. Early diagnosis is important for emergency surgical treatment.

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**A**neurysms of sinus of Valsalva (ASV) are rare cardiac abnormalities. They are dilatations of the aortic sinuses and are classified as congenital or acquired. Sometimes they coexist with other congenital heart diseases and they often rupture into the heart chambers causing acute clinical syndrome. ASVs and coexisting congenital heart disease can easily be diagnosed with echocardiography. Early diagnosis and immediate surgical treatment can save the patient's life in most cases. At present, the surgical correction of the rupture has a very low perioperative risk and satisfactory long-term therapeutic results.

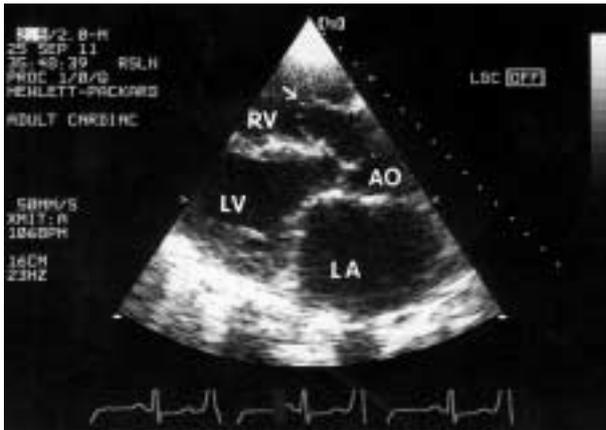
A case of a ruptured aneurysm of the right sinus of Valsalva into the right ventricle is presented below. The usefulness of various diagnostic methods, as well as the surgical treatment are also discussed.

### Case description

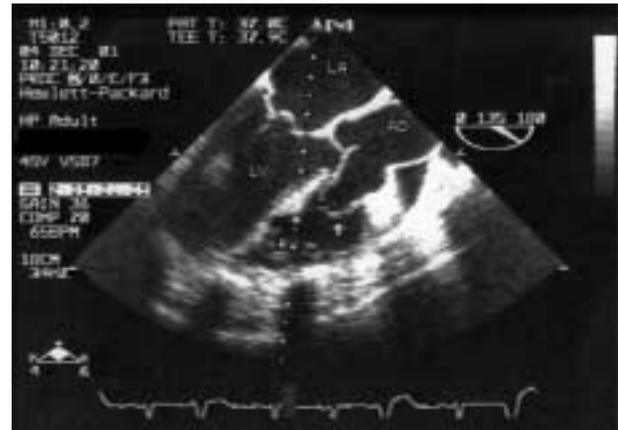
A 42-year old man was admitted to our Department because of palpitations and

exertional dyspnea-orthopnea, which progressively worsened during a 20-day period. On physical examination, he had a heart rate of 110 beats/min, a blood pressure of 100/40 mmHg and was feverless. On auscultation, there was a continuous murmur along the left parasternal line, accompanied by a thrill. The ECG showed sinus rhythm with frequent ventricular premature beats (bigeminy), while the chest X-ray showed an increase of the cardiothoracic index (21/32) and periportal pulmonary oedema. The blood test results were within normal limits.

The transthoracic echocardiogram showed small increase in the dimensions of the left ventricle with normal systolic function, prolapse of the posterior mitral leaflet with slight regurgitation of the valve and aneurysmal dilatation of the right sinus of Valsalva protruding into the right ventricle (Figure 1). The transesophageal echocardiogram confirmed the above mentioned findings (Figure 2), while in the color Doppler, a turbulent flow was observed from the aorta to the right ventri-



**Figure 1.** Parasternal long axis view at transthoracic echocardiography. Aneurysmal dilatation (arrow) of right sinus of Valsalva protruding into the right ventricle (RV). (AO: aorta, LA: left atrium, LV: left ventricle).



**Figure 2.** Modified transesophageal long axis view of the ascending aorta in 143° demonstrating the aneurysm of the right sinus of Valsalva (arrow) protruding into the right ventricle (RV). (AO: aorta, LA: left atrium, LV: left ventricle).

cle, through the ruptured aneurysm (Figure 3). Finally, a systolic-diastolic flow was recorded in the continuous Doppler, due to the fact that the pressure in the aorta was higher than the pressure in the right ventricle throughout the cardiac cycle.

The patient, without prior angiography, was subjected to surgery in order to repair the ASV rupture with a patch.

## Discussion

ASVs are classified as congenital or acquired. Congenital ASVs are caused by separation or failed fusion of the aortic media with the fibrous annulus of the aortic valve. Acquired ASVs can develop as the result of injury<sup>1</sup>, endocarditis<sup>2</sup>, syphilis<sup>3</sup>, Behcet's disease<sup>4</sup> and Marfan syndrome. Isolated ASVs rarely occur in Marfan syndrome and the embryogenic association of the two conditions is disputed<sup>5</sup>.

The aneurysms are primarily located in the right sinus of Valsalva (76.8%) and secondary in the non-coronary sinus (20.2%) and in the left sinus (3%)<sup>6</sup>. The most common coexisting congenital heart diseases are ventricular septal defect (VSD), usually subaortic (25-55%) and regurgitation of the aortic valve and rarely pulmonary stenosis, patent ductus arteriosus, atrial septal defect, subaortic stenosis and tetralogy of Fallot<sup>7</sup>.

ASVs can remain silent for several years and usually rupture during the 3rd or 4th decade of life into the heart chambers (usually in the right chambers)<sup>7</sup>, into the interventricular septum (causing complete atrioventricular block)<sup>8</sup> or in the pericardial

cavity<sup>9</sup>. Initially, a rupture in an endocardial chamber can be silent but later manifests as progressive heart failure, caused by arteriovenous communication and aortic regurgitation<sup>10</sup>. There is no clear correlation in the literature between physical/athletic activities and the time of rupture of the ASVs<sup>11</sup>.

Unruptured ASVs can cause obstruction of the right ventricular outflow, infectious endocarditis, thrombus formation with subsequent embolic events<sup>7</sup> and myocardial ischemia/ infarction, due to compression of the stem or its main branches by the body of the aneurysm of the left sinus or obstruction of the ostium of the right coronary artery by thrombus and/or syphilis of the aneurysm of the right sinus of Valsalva<sup>12</sup>.



**Figure 3.** Transesophageal color Doppler recording in the modified short axis view showing shunt flow (black arrow) from the aorta (AO) to the right ventricle (RV). (LA: left atrium, RA: right atrium).

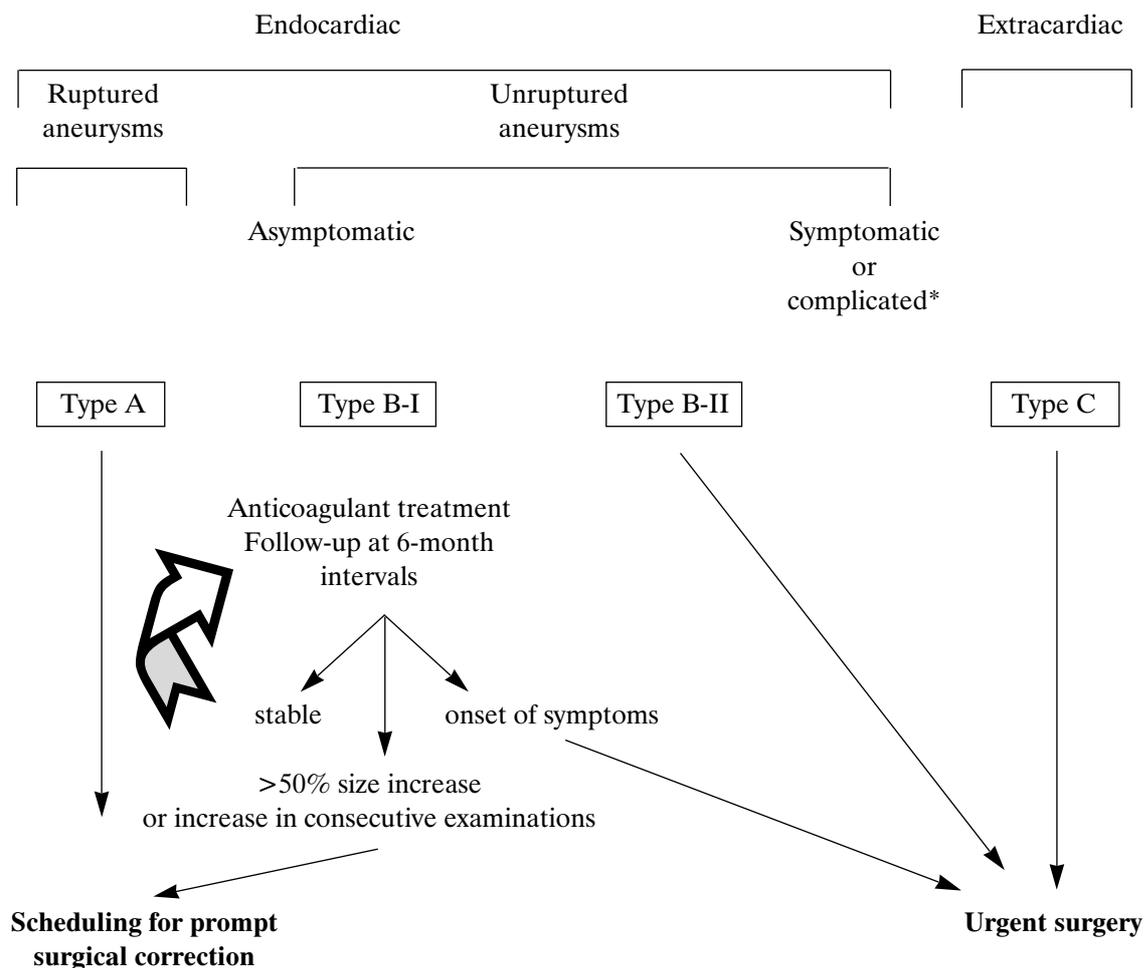
Transthoracic echocardiography is the main diagnostic tool for the identification of the ASVs<sup>13</sup>, while transesophageal imaging may further help the diagnosis and the cardio-surgical correction of the lesion, especially when VSD is also present<sup>14</sup>. Echocardiography can also provide important information necessary for the differential diagnosis from other disorders which cause continuous murmur, such as patent ductus arteriosus, aorticopulmonary window, coronary fistula, as well as coexisting VSD and aortic valve regurgitation.

Hemodynamic and angiographic study, although limited by the echocardiographic examination, is used to detect the site of rupture as well as to estimate the patency of the coronary vessels, particularly in elderly patients. Finally, magnetic resonance imaging may be used to confirm the diagnosis<sup>15</sup>.

Sakakibara and Konno<sup>16</sup> in an effort to describe the initial position of the aneurysmal sac, the direction of the dilatation, the heart chamber in which the

rupture takes place and the combined lesions, classified congenital ASVs into 4 categories (Table 1). ASV which do not match the typical anatomic models of the 4 categories are in most cases acquired (endocarditis, syphilis, injury).

Vural et al<sup>17</sup> used a simple and functional classification of the therapeutic approach to ASVs, based on the clinical picture and the echocardiographic findings (Figure 4). Surgical intervention is required both for ruptured aneurysms and for symptomatic unruptured ones. As far as unruptured asymptomatic aneurysms are concerned, their size determines whether surgical treatment is required or not. Surgical treatment is necessary, if the size of the aneurysm is larger than 50% of the average size of the other two normal Valsalva sinuses or is increasing in consecutive echocardiographic examinations. In addition, patients should be operated on if there is compression or malformation of the adjacent tissues.



**Figure 4.** The algorithm for therapeutic approach to ASV, by Vural et al<sup>17</sup>.

\* e.g. occlusion of the right ventricular outflow tract, aortic valve regurgitation, ASV: Aneurysm of sinus of Valsalva.

**Table 1:** Classification system of ASV by Sakakibara and Konno<sup>16</sup>.

**Type I:** the aneurysm originates in the left portion of the right sinus, protrudes forward and ruptures into the right ventricle near the pulmonary valve. The concurrent presence of VSD under the pulmonary valve is frequent.

**Type II:** the aneurysm originates in the mid portion of the right sinus, protrudes and ruptures in the right ventricle. A concurrent VSD is uncommon.

**Type III:** the aneurysm originates in the mid portion of the right coronary leaflet and protrudes towards the tricuspid valve. It often ruptures into the right atrium and sometimes into the right ventricle, just below the septal leaflet of the tricuspid valve. VSD is rarely encountered.

**Type IV:** the aneurysm originates in the right portion of the non-coronary leaflet and ruptures into the right atrium. A combined VSD is uncommon.

ASV: Aneurysm of sinus of Valsalva.

VSD: Ventricular septal defect.

Therapeutic treatment includes surgical methods such as direct suturing or suturing with the use of a patch, as well as the defect closure by intravenous placement of an occluder<sup>18</sup>. According to Ghosh et al<sup>19</sup> in cases of both extensive coronary artery disease and unruptured ASV, a simultaneous bypass and correction of the aneurysm should be preferred, due to the unknown physical history of the latter and the risk of rupture and complications.

Surgical treatment presents low operative risk and has a high long-term survival rate, almost similar to that of healthy individuals when there are no concurrent congenital malformations. Mayer et al<sup>11</sup> and Lukacs et al<sup>20</sup> report a zero perioperative mortality in a total of 30 patients with ruptured ASV and advise early intervention. The largest series in the literature includes 129<sup>21</sup> patients, out of whom 50% presented with ruptured ASV. The perioperative mortality was 4%, due to a pre-existing endocarditis-sepsis in 3/4 of the cases. An early surgical intervention should therefore be encouraged since the perioperative risk is low and as it halts disease progress and prevents a delayed, complicated and less satisfactory correction.

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