Aortic Sinus of Valsalva Aneurysms Simulating Primary Right-sided Valvular Heart Disease

By Bernadine H. Bulkley, M.D., Grover M. Hutchins, M.D., and Richard S. Ross, M.D.

SUMMARY
While acute aortic regurgitation is the most common valvular lesion associated with aortic sinus of Valsalva aneurysms, this report describes the pathological findings in five patients in whom primary right-sided valve dysfunction was simulated by sinus of Valsalva aneurysms. In two patients aneurysms of the noncoronary sinus projected into the right atrium at the level of the tricuspid valve and caused tricuspid incompetence. In three patients aneurysms of the right sinus of Valsalva projected into the right ventricle immediately below the pulmonic valve and caused pulmonary outflow tract obstruction. In only one of these patients was the aneurysm perforated at necropsy. Although valvular dysfunction was evident clinically, in none was there a clinical suspicion of a sinus of Valsalva aneurysm. In addition to valvular dysfunction, two patients had conduction disturbances and two, right coronary arterial occlusions found to be the result of aneurysms. The risks of valve dysfunction, arrhythmias, aneurysm rupture, and sudden death and the ability to correct this lesion surgically make it especially important to consider aortic sinus of Valsalva aneurysm as a cause of obscure right-sided valvular disease.

AORTIC ANEURYSMS of the sinuses of Valsalva are not usually clinically apparent unless perforation occurs, which simulates acute aortic regurgitation. There are a few reported cases in which unperforated sinuses of Valsalva aneurysms caused conduction disturbances or myocardial ischemia due to coronary arterial occlusion, but the finding of this lesion in the presence of right-sided valvular dysfunction is rare. This report describes five necropsy patients in whom tricuspid incompetence or pulmonary stenosis was caused by sinus of Valsalva aneurysms. In only one of these patients had the aneurysm ruptured.

Patients Studied and Findings
Of 31 patients with sinus of Valsalva aneurysms in the autopsy files of The Johns Hopkins Hospital, five had aneurysms which impaired the flow of blood in the right side of the heart (table 1). The five patients ranged in age from 14–62 years, and four were men. A history of syphilis was present in three (cases 3, 4, 5). During life, tricuspid regurgitation was diagnosed in two patients (cases 1, 2) and pulmonic stenosis in one (case 3). In the remaining patients, a systolic murmur and loud P2 were described in case 4, and a murmur of aortic regurgitation associated with a history of syphilis, in case 5. Electrocardiograms, available in two patients (cases 1, 2), showed first degree and left bundle branch block and premature ventricular contractions in one patient, and right bundle branch block and complete atrioventricular block in the other. An echocardiogram in the latter patient showed dilatation of the aortic root and both ventricles, and these findings, along with the phonogram, were interpreted as compatible with tricuspid regurgitation. None of the patients underwent cardiac catheterization, and the diagnosis of sinus of Valsalva aneurysm was not suggested clinically in any of them. Death was sudden and unexpected in four patients, and a consequence of clinically unexplained intractable right-sided heart failure in the other.

At necropsy three patients had unruptured sinus of Valsalva aneurysms involving the right coronary sinus and projecting into the right ventricular outflow tract (fig. 1) (cases 3, 4, and 5). The aneurysms measured up to 4 cm in diameter and each produced narrowing of the outflow tract of the right ventricle of greater than 50%. In two of these patients the pulmonary valves were normal, but in one (case 3), the aneurysm projected between two cusps of the pulmonic valve and caused retraction of one cusp which may have caused pulmonic insufficiency although a murmur of pulmonic insufficiency was not detected. Right ventricular hypertrophy was present in all three hearts. Occlusion of the right coronary orifice by a thrombus in the sinus of Valsalva aneurysm occurred in two
Table 1

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<th>Patients Studied</th>
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<tr>
<td><strong>Tricuspid Regurgitation</strong></td>
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<td>1 62 M</td>
<td>3/6 pansystolic murmur left lower sternal border (LLSB); hepatomegaly; ankle edema; ECG-RBBB, A-V block; echocardiogram-dilatation right ventricle, paradoxical septal motion. Died suddenly.</td>
<td>Posterior S of V aneurysm projecting into RA with perforation; RVH; healed myocardial infarct. Severe chronic passive congestion of liver.</td>
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<td>2 14 F</td>
<td>3/6 systolic murmur LLSB; pulsatile liver, ankle edema; ECG-LBBB, 1st heart block, PVC's. Died with intractable congestive heart failure.</td>
<td>Posterior S of V aneurysm projecting into RA at level of tricuspid valve deforming septal leaflet. Severe chronic passive congestion of liver.</td>
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<td><strong>Pulmonary Outflow Tract Obstruction</strong></td>
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<td>3 37 M</td>
<td>4/6 systolic murmur pulmonic area, hepatomegaly; clinical diagnosis of congenital pulmonic stenosis; positive serology for syphilis. Died suddenly.</td>
<td>Right S of V aneurysm projecting into pulmonary outflow tract (POFT) and between 2 cusps of pulmonic valve; occlusion RCA ostium by thrombus in aneurysm. Syphilitic aortitis. Severe chronic passive congestion of liver.</td>
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<tr>
<td>4 37 M</td>
<td>3/6 systolic murmur at base; loud P2; hepatomegaly; positive serology for syphilis. Died suddenly.</td>
<td>Right S of V aneurysm projecting into POFT; RVH. Syphilitis aortitis. Severe chronic passive congestion of liver.</td>
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<tr>
<td>5 33 M</td>
<td>2/6 systolic murmur at base; 3/6 diastolic blow; orthopnea; ankle edema; positive serology for syphilis. Died suddenly.</td>
<td>Right S of V aneurysm (3.5 X 5 X 4 cm) projecting into POFT; RVH; occlusion RCA ostium by thrombus within aneurysm; acute myocardial infarction. Syphilitic aortitis with aortic valve incompetence. Severe chronic passive congestion of liver.</td>
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Abbreviations: A-V = atrioventricular; ECG = electrocardiogram; LBBB = left bundle branch block; PVC = premature ventricular contraction; RBBB = right bundle branch block; RCA = right coronary artery; RVH = right ventricular hypertrophy; S of V = Sinus of Valsalva.

patients, both of whom died suddenly (cases 3 and 5). In two patients a sinus of Valsalva aneurysm of the posterior or noncoronary sinus projected into the right atrium immediately above the tricuspid valve, immobilized a portion of the septal tricuspid leaflet and rendered the valve incompetent. In case 1, the aneurysm had a small perforation (fig. 2), but there had been no continuous murmur or evidence of a right to left shunt noted clinically.

Three of the aneurysms were syphilitic in origin. Each of these patients had positive serologic tests for syphilis and histologic evidence of syphilitic aortitis at autopsy. Aortic regurgitation due to syphilis was also present in case 5. The etiology of the aneurysms in the other two patients was uncertain: in case 1 a history of alcoholism and multiple traumatic injuries suggested a traumatic cause for the aneurysm; in case 2 the aneurysm was interpreted as a healed valve ring abscess.

**Discussion**

Aortic aneurysms of the sinuses of Valsalva involve the right or noncoronary sinus in 90% of instances and frequently bulge into a cardiac chamber. Patients with these lesions have been found to be asymptomatic clinically unless the aneurysm ruptures. 1, 2, 8, 9 In contrast, sinus of Valsalva aneurysms caused pulmonary outflow obstruction in three of our patients and tricuspid regurgitation in two, and in only one of the five patients had the aneurysm ruptured. A clinical diagnosis of tricuspid or pulmonic disease was made in three patients but in none was a sinus of Valsalva aneurysm suspected. Pathologic evidence for functionally significant right-sided cardiac lesions included right ventricular hypertrophy and severe chronic passive congestion of the liver in all cases.

Aneurysms of the right sinus of Valsalva most often project into the right ventricle, and those of the noncoronary and posterior sinus, into the right atrium. 10, 11 The anatomic relationship between the aortic sinus of Valsalva and the cardiac chambers explains these relationships and the physiologic consequences of both ruptured and unruptured aneurysms (table 2). At the level of the posterior coronary cusp, the aorta abuts on the right atrial wall immediately above the septal tricuspid leaflet (fig. 3). Aneurysms in this posterior sinus project into the adjacent right atrium and may cause distortion or destruction of the septal
portion of the tricuspid valve with attendant tricuspid incompetence. Hence, noncoronary sinus of Valsalva aneurysms may rupture into the right atrium or into the most cephalad portion of the right ventricle at the level of the tricuspid valve.

The right coronary sinus is adjacent to the pulmonary outflow tract immediately below the pulmonic valve (fig. 3). Aneurysms of the right sinus therefore protrude into the pulmonary outflow tract and almost always rupture into the right ventricle; in rare cases they may be high enough to rupture into the pulmonary trunk. A right sinus of Valsalva aneurysm projecting into the right ventricle below the pulmonic valve may cause pulmonary outflow tract obstruction, as in our three patients with large unruptured right sinus aneurysms. Pulmonary insufficiency may also have resulted from distortion of the pulmonary leaflets by the aneurysm in one patient. Although subpulmonic stenosis resulting from unruptured sinus of Valsalva aneurysms is a rare finding, Seymour et al. determined that aneurysms of the ascending aorta, usually luetic, compressing the right ventricular outflow tract, were the most common cause of acquired pulmonary stenosis.

Complete heart block and A-V nodal rhythms described in patients with sinus of Valsalva aneurysms have been attributed to A-V node or His bundle injury. Two of our patients had conduction disturbances, including first degree, bundle branch, and complete atrioventricular block, and it is likely that protrusion of the sinus aneurysms through the cephalad portion of the septum led to these disturbances. In case 1 episodes of complete atrioventricular block were unexplained clinically, and the sudden and unexpected death was presumed to be caused by a conduction disturbance.

Sudden death also occurred in two other patients (cases 3, 5) and obstruction of the ostia of the right coronary artery by thrombus within the right sinus of Valsalva aneurysm was found at autopsy. In both patients, who were in their mid-thirties in age, these coronary occlusions appeared the most likely explanation for their sudden demise. Eliot et al. have described coronary insufficiency in a 54-year-old woman due to compression of the left coronary artery by a rare left sinus of Valsalva aneurysm.

In retrospect, the only clinical evidence of the sinus aneurysms in our patients was suggestive of primary
tricuspid or pulmonic valvular disease and led to mistaken diagnoses. Unfortunately, none of these patients underwent cardiac catheterization. Sinus of Valsalva aneurysms would have been evident by cardiac angiography and well might have been surgically repaired. The hazards of valvular dysfunction, aneurysm rupture, arrhythmias and sudden death from a potentially correctable lesion make it especially important to consider the possibility of an unruptured aortic sinus of Valsalva aneurysm as the primary lesion in patients presenting with obscure right-sided valvular disease.

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B H Bulkley, G M Hutchins and R S Ross

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