CASE REPORT

Echocardiographic Manifestation of Right Sinus of Valsalva Aneurysm

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SUMMARY
This case report describes the distinctive echocardiographic pattern in a patient with a congenital right sinus of Valsalva aneurysm. The echocardiogram demonstrated an abnormal structure extending from the aortic root into the left ventricular chamber adjacent to the interventricular septum. The structure showed diastolic motion away from the septum and systolic motion toward the septum. This movement was consistent with cineangiographic filling of the aneurysm during diastole and emptying during systole. After operative closure of the aneurysm, the echocardiogram showed dense thickening along the aortic root and contiguous interventricular septum, probably representing thrombosis and fibrosis within the obliterated aneurysmal sac; but no abnormal movement of the structure was present.

Additional Indexing Words:
Bicuspid aortic valve Aortic valve echocardiogram Aortic insufficiency Aortic stenosis Ultrasound

SINCE EDLER'S1 original echocardiographic description of the aortic valve, abnormal aortic valve echo patterns have been described in such conditions as valvular aortic stenosis,2 hypertrophic3 and discrete4 subaortic stenosis, truncus arteriosus and low cardiac output,5 and endocarditis.6 In the present report we describe a patient with clinical features of aortic stenosis and insufficiency who had a distinctive echocardiographic structure within the aorta and base of the left ventricle which at cardiac catheterization was found to be a right sinus of Valsalva aneurysm protruding into the left ventricle. After surgical closure of the aneurysm, the region of the left ventricular outflow tract and aortic root remained thickened; but the abnormal preoperative echocardiographic motion of the aneurysm was no longer present.

Case Report
A 31-year-old male was first noted to have the murmurs of aortic stenosis and insufficiency at age 22. During a routine cardiac clinic visit, an unusual echocardiographic pattern was noted in the aortic valve region; and the patient was admitted to the Indiana University Medical Center for further evaluation. Symptoms at the time of admission included exertional dyspnea, dizziness and palpitations. He denied chest pain, fever, chills or weight loss.

On physical examination the pulse was 80 and the blood pressure was 115/50 mm Hg. The jugular venous pressure was not elevated. The carotid upstroke was brisk with bisferiens quality. The lungs were clear to auscultation and percussion. On cardiac examination the patient had an enlarged heart with the P1 in the anterior axillary line in the sixth left intercostal space. There was a prominent left ventricular lift and a systolic thrill along the left sternal border. The first heart sound was normal, the second heart sound was paradoxically split. There was an ejection click followed by a grade 4/6 systolic ejection murmur which was loudest in the aortic area and radiated to the carotids. There was a grade 4/6 prolonged diastolic decrescendo murmur along the left sternal border. A fourth heart sound was present. The remainder of the physical examination was normal. The electrocardiogram showed...
left bundle branch block; the chest X-ray showed marked left ventricular enlargement. The echocardiogram revealed an abnormal structure extending from the aortic root into the left ventricular chamber adjacent to the interventricular septum (fig. 1). During diastole the structure moved away from the septum and toward the anterior mitral valve leaflet. During systole the structure moved toward the septum and into the aortic root in the position normally occupied by the right aortic valve leaflet. Also in the echocardiogram there was marked left ventricular dilatation and abnormal septal motion characteristic of left bundle branch block.7 Fluttering of the anterior mitral valve leaflet was noted, indicative of aortic insufficiency. Cineangiography demonstrated a right sinus of Valsalva aneurysm protruding into the left ventricle along the interventricular septum (fig. 2). The diastolic filling and systolic emptying of the aneurysm corresponded to the diastolic and systolic motion of the abnormal structure on the echocardiogram. Aortic stenosis and insufficiency were present (table 1).

On physical examination there were the following changes: the systolic ejection click was no longer sinus tract in the right aortic leaflet (fig. 3). Two months postoperatively, the patient was re-evaluated.

The patient underwent surgical closure of the aneurysmal sac by the placement of a patch over the present; both the systolic and diastolic murmurs were decreased in intensity, and the diastolic murmur was shortened in duration. The echocardiogram demonstrated dense thickening along the aortic root and contiguous interventricular septum; however the septum became thinner as the ultrasonic beam was directed deeper into the left ventricular cavity (fig. 4). Since the aneurysm was not excised but merely closed, the dense echoes probably represented thrombosis and fibrosis within the aneurysmal sac. Cineangiography revealed deformity of the right sinus of Valsalva in the area of the patch graft but no evidence of aneurysmal filling (fig. 5). There was a decrease in both the aortic stenosis and insufficiency (table 1).

Discussion

Aneurysms of the sinus of Valsalva may be either congenital or secondary to such underlying diseases

<table>
<thead>
<tr>
<th>Hemodynamic Data</th>
<th>Preoperative</th>
<th>Postoperative</th>
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<tbody>
<tr>
<td>HR (beats/min)</td>
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<tr>
<td>RA (mm Hg)</td>
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<td>3/3* (1)</td>
</tr>
<tr>
<td>PA (mm Hg)</td>
<td>35/20 (26)</td>
<td>23/10 (14)</td>
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<td>PCW (mm Hg)</td>
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<tr>
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<td>CI (l/min/m²)</td>
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<td>23</td>
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<tr>
<td>AI</td>
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<td>Mild</td>
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</table>

Abbreviations: HR = heart rate; RA = right atrial pressure; PA = pulmonary artery pressure; PCW = pulmonary capillary wedge pressure; LV = left ventricular pressure; AA = ascending aortic pressure; CI = cardiac index; AVG = aortic valve gradient; AI = aortic insufficiency.

*"a" and "v" wave pressures respectively.
†Mean pressures in parentheses.

Figure 1
Echocardiogram of patient with right sinus of Valsalva aneurysm. The anterior aortic valve echo (AV) demonstrates exaggerated anterior systolic motion with eccentricity of the aortic valve leaflets. Although this abnormal echo occupies the position of the normal right aortic valve cusp in the aortic area, it protrudes (arrows) into the left ventricular cavity (LV). With the onset of diastole the echo moves rapidly away from the interventricular septum, only to return toward the septum with systole. LA = left atrium; MV = mitral valve.

Figure 2
Left anterior oblique cineangiogram of patient with right sinus of Valsalva aneurysm. A. Diastole. Contrast-filled aneurysm (arrows) projects from the aortic root into the left ventricular cavity along the septal margin. B. Systole. Aneurysm empties of contrast medium. Three aortic sinuses are seen.
as syphilis, bacterial endocarditis, atherosclerosis, and aortic dissection. As our patient had no findings to support an acquired etiology, his aneurysm was considered to be congenital in origin. In congenital sinus of Valsalva aneurysm the basic defect is believed to be in the development of the annulus fibrosis. As a result of the failure of fusion between the bulbar septum and the aorto-pulmonary septum at the annulus fibrosis, the high intraluminal aortic pressure eventually causes aneurysmal dilatation of the sinus of Valsalva at the site of congenital weakness. Congenital sinus aneurysms are confined almost exclusively to the right coronary sinus. The ventricular septum lies below all parts of this sinus. Aneurysmal dilatation of the right sinus usually results in herniation or perforation into the right ventricle. Occasionally, the aneurysm prolapses into the left ventricle; the

displacement of the associated, non-supported right aortic valve leaflet often results in aortic incompetence. Because of the usual location of an unruptured right sinus of Valsalva aneurysm in the outflow tract of the right or left ventricle, the clinical findings characteristically mimic aortic valvular disease. Our patient was typical in this regard; he presented with murmurs of aortic stenosis and insufficiency. The echocardiogram, however, delineated the aneurysm and its anatomic relation to surrounding structures. The aneurysm extended from the aortic root into the left ventricle; it filled during diastole and emptied during systole. Since the right aortic cusp exhibits anterior systolic motion in the aortic root, the presence of the aneurysmal echo in this location would imply an origin from the right sinus of Valsalva. Also the herniation of the aneurysm into the left ventricular cavity adjacent to the interventricular septum would be most compatible anatomically with a right sinus of Valsalva aneurysm. In our experience the only other entity which is associated with a similar echocardiographic pattern in the aorta is a congenitally bicuspid aortic valve (fig. 6). However, the abnormal echoes are confined exclusively to the aortic root in the case of a congenitally bicuspid aortic valve, whereas the abnormal echo extended well into the left ventricular cavity in this patient with a sinus of Valsalva aneurysm (fig. 1).

Although right ventricular outflow tract obstruction has been reported due to an unruptured sinus of Valsalva aneurysm, significant left ventricular outflow obstruction has not been previously described. Preoperatively the aortic valve gradient was 46 mm Hg; postoperatively it was 23 mm Hg. The decrease in the aortic valve gradient was felt to be a result of the elimination of the left ventricular outflow obstruction produced by the aneurysmal sac. Also the patch graft gave added support to the right aortic valve leaflet, thereby decreasing the amount of aortic valve prolapse and insufficiency.

Of clinical importance, the abnormal echocardiographic features of this patient included a markedly deformed right sinus of Valsalva aneurysm. The right aortic sinus was somewhat deformed as a result of surgery.

Figure 4
Echocardiogram of patient after successful surgical closure of right sinus of Valsalva aneurysm. The anterior aortic wall and contiguous interventricular septum (IVS) are thickened in the area where the sinus of Valsalva aneurysm was previously seen. The septum is of normal thickness near the apex. LA = left atrium; LV = left ventricular cavity; MV = mitral valve; AO = aorta.

Figure 5
Left anterior oblique cineangiogram of patient after surgical closure of right sinus of Valsalva aneurysm. In diastole there is no evidence of the aneurysm. The right aortic sinus is somewhat deformed as a result of surgery.

Figure 6
graphic pattern in this patient demonstrated that this was not a case of uncomplicated aortic valvular disease. Although this echocardiographic pattern may not always be present in unruptured sinus of Valsalva aneurysm, the presence of such an abnormal echo-producing structure should suggest this lesion.

Acknowledgment
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References
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