Echocardiographic Diagnosis of Congenital Sinus of Valsalva Aneurysm with Dissection Of the Interventricular Septum

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SUMMARY A case is reported in which a congenital aneurysm of the right coronary sinus of Valsalva ruptured and dissected into the interventricular septum. M-mode and cross-sectional echocardiographic examination allowed accurate preoperative assessment of the pathologic anatomy, which was confirmed by angiography, surgery and autopsy. Dissection of the interventricular septum by a congenital sinus of Valsalva aneurysm is a rare lesion, has a poor prognosis, and can be diagnosed noninvasively with echocardiography.

CONGENITAL ANEURYSM of the sinus of Valsalva is frequently encountered in clinical practice. Recent studies have emphasized the wide variety of clinical manifestations of this lesion, depending on its location and course. In this report we present a case in which a ruptured right coronary sinus of Valsalva aneurysm took an unusual course, dissecting into the muscular interventricular septum. We believe this to be the first report of the preoperative diagnosis of this condition.

Case Report

A 16-year-old black male presented to the University of Cincinnati Hospital emergency room for evaluation of left-sided chest pain related to slight trauma that occurred 2 days before. Physical examination revealed systolic and diastolic murmurs, and an ECG revealed complete left bundle branch block. A chest x-ray was interpreted as normal. The patient was referred for further cardiac evaluation.

The patient was the product of a normal pregnancy, labor and delivery with no history of perinatal cyanosis or respiratory difficulties. His subsequent growth and development were unremarkable. There was no history consistent with rheumatic fever or heart disease. Specifically, there had been no episodes of chest pain, dizziness, dyspnea, or syncope. A complete physical examination several years before reportedly revealed no cardiac abnormalities. He was active in competitive athletics. There was no history of abuse of controlled substances. Eight siblings were all in good health, and there was no family history suggestive of Marfan’s syndrome.

The findings of the initial diagnostic evaluation in May 1979 were consistent with severe aortic regurgitation. Cardiac catheterization was refused. During the ensuing 6 months, the patient developed vague chest pain and progressive exertional dyspnea. Two weeks before admission, the patient developed increasing orthopnea and a nocturnal nonproductive cough. One week later, dyspnea increased, accompanied by several episodes of syncope. He was admitted to the University of Cincinnati Hospital, Cincinnati General Division in November 1979 for evaluation of congestive heart failure.

Physical examination revealed a tall, slender adolescent black male in mild respiratory distress. There were no stigmata of Marfan’s syndrome. The blood pressure was 110/50 mm Hg in both arms. The heart rate was 110 beats/min and cervical venous pressure was normal. The carotid upstrokes were brisk, with rapid emptying. Auscultation of the lungs revealed bibasilar end-inspiratory rales. On cardiac examination, the apical impulse was in the fifth intercostal space 1 cm lateral to the midclavicular line. Prominent apical and parasternal lifts were palpable. In addition, there was a diastolic thrill along the left sternal border. The first heart sound was decreased in intensity; the second heart sound was single. There was a grade III/VI systolic ejection murmur at the base that radiated to the neck, and a grade III/VI holosystolic blowing murmur at the apex that radiated to the left axilla. A loud, harsh diastolic decrescendo murmur along the left sternal border was best heard in the third left intercostal space. Apical S₃ and S₄ were audible and palpable. The remainder of the examination was not remarkable.

Initial laboratory studies, including complete blood count, serum electrolytes, blood urea nitrogen and creatinine, serum glucose, liver function tests, coagulation studies, and serologic tests for syphilis were normal. The chest roentgenogram demonstrated mild cardiomegaly without specific chamber enlargement. There were early signs of congestive cardiac
failure. The ECG again revealed complete left bundle branch block and abnormal left-axis deviation.

Echocardiogram

Figure 1 is a representative sweep from aorta to left ventricle from the preoperative M-mode echocardiogram. The left ventricle appeared dilated and systolic contraction was decreased. There was no diastolic fluttering of the anterior mitral leaflet. At approximately the level of the anterior mitral leaflet, echoes from the intraventricular septum separated into two distinct echoes approximately 1.5 cm apart that moved in parallel. These two echoes diverged at the level of the aortic valve to become continuous with a large echo-free space anterior to the aortic valve. Abnormal echoes associated with the aortic valve were present during diastole in the left ventricular outflow tract, and were presumed to represent prolapse of some portion of the aortic valve during diastole. Cross-sectional echocardiography revealed a large aneurysm of the right coronary sinus of Valsalva (fig. 2). A defect was seen in the aortic wall through which portions of the aortic valve crossed during systole, with prolapse of these structures toward the left ventricular outflow tract during diastole. The long-axis

![Echocardiogram](image-url)
view of the left ventricle and mitral valve (fig. 3) revealed a longitudinal defect in the interventricular septum that was continuous with the aortic sinus aneurysm and extended into the muscular portion of the interventricular septum at least to the level of the body of the mitral leaflets.

Cardiac Catheterization

Therapy was begun with digoxin and diuretics. One day later, the ECG revealed complete atrioventricular block and accelerated idioventricular rhythm. The serum digoxin level was within the therapeutic range. On November 19, 1979 cardiac catheterization was performed. The hemodynamic data are listed in table 1. Catheterization of the right heart revealed moderately elevated pulmonary arterial and right ventricular systolic pressures. There was no evidence of left-to-right intracardiac shunting. The left ventricle was markedly dilated and systolic function was diminished. Aortography (fig. 4) revealed severe aortic regurgitation. There was immediate opacification of an area corresponding to the upper portion of the interventricular septum. This “septal chamber” filled during late systole and early diastole and narrowed somewhat during ventricular ejection. There was no opacification of right-heart chambers.

Surgical Findings

The patient underwent surgery on November 24, 1979 through a median sternotomy. The left ventricle was markedly enlarged, with severe hypertrophy and decreased contractile function. There was a defect measuring 1.0 \( \times \) 0.8 cm distal to the attachment of the right coronary cusp and between the cusp attachment and the orifice of the right coronary artery, extending into the middle of the interventricular septum. At the septal extent of this cavity, a large windsock defect extended into the left ventricle and terminated in an 8-mm perforation. The majority of the rapid aortic

![Figure 3. Long-axis presentation of mitral valve, aorta (Ao) and left ventricle (LV) from preoperative cross-sectional echocardiogram. A and B (top) represent caudal and cephalad orientation of the transducer, respectively, in the same examining plane. The line drawing (bottom) reconstructs the entire long-axis view. Note the extension of the aortic sinus aneurysm (An) into the interventricular septum, and diastolic prolapse of the damaged aortic cusps (arrow) into the left ventricular outflow tract. RV = right ventricle; LA = left atrium.](image-url)

<table>
<thead>
<tr>
<th>Table 1. Hemodynamic Data</th>
<th>Pressure (mm Hg)</th>
<th>Oxygen saturation (%)</th>
<th>Cardiac output*</th>
<th>Cardiac index</th>
<th>Stroke volume (forward)</th>
<th>Stroke volume (total)</th>
<th>Stroke volume (regurgitant)</th>
<th>Ejection fraction</th>
<th>Regurgitant fraction</th>
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<tbody>
<tr>
<td>Right atrium</td>
<td>3</td>
<td>60</td>
<td>Cardiac output*</td>
<td>3.74 l/min</td>
<td></td>
<td></td>
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<tr>
<td>Right ventricle</td>
<td>42/3</td>
<td>60</td>
<td>Cardiac index</td>
<td>2.78 l/min</td>
<td>Stroke volume (forward)</td>
<td>37.4 ml/beat</td>
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<tr>
<td>Pulmonary artery</td>
<td>42/27; 33</td>
<td>62</td>
<td>Stroke volume (forward)</td>
<td>37.4 ml/beat</td>
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<td>Pulmonary wedge</td>
<td>26</td>
<td></td>
<td>End-diastolic volume†</td>
<td>375.1 ml</td>
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<td></td>
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<tr>
<td>Left ventricle</td>
<td>85/26</td>
<td>99</td>
<td>End-systolic volume†</td>
<td>165.4 ml</td>
<td>Stroke volume (total)</td>
<td>209.7 ml</td>
<td>Stroke volume (regurgitant)</td>
<td>171.7 ml</td>
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<tr>
<td>Aorta</td>
<td>85/50; 62</td>
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<td>Stroke volume (total)</td>
<td>209.7 ml</td>
<td>Stroke volume (regurgitant)</td>
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<td>Body surface area</td>
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<td>Ejection fraction</td>
<td>44%</td>
<td>Regurgitant fraction</td>
<td>45%</td>
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*Thermodilution.
†Kennedy-Dodge (right anterior oblique).
runoff was through the aneurysm of the sinus of Valsalva, and not through the aortic valve. The left and noncoronary cusps were normal, but the right coronary cusp was shortened and deformed, necessitating aortic valve replacement. The aneurysm of the right sinus of Valsalva was excised, and the defect was repaired parallel to the attachment of the cusp to close off the left ventricular outflow tract from the opening into the ventricular septum and into the aortic root. A 23-mm Björk-Shiley aortic prosthesis was sewn into place.

Course

Postoperatively, a permanent pulse generator was implanted due to intermittent second-degree atrioventricular block. During stable convalescence, an episode of ventricular fibrillation, presumed to be related to quinidine, occurred without sequelae. Six weeks postoperatively, evidence of mild congestive heart failure was present, and the patient was begun on digoxin and furosemide, with some symptomatic improvement. The postoperative echocardiogram (fig. 5) revealed resolution of the previously noted septal abnormality. Symptoms of congestive cardiac failure returned 10 weeks after operation; therapy with hydralazine and isosorbide dinitrate resulted in hemodynamic and clinical improvement. Two weeks later (3 months after surgery) the patient developed ventricular fibrillation while in the hospital. Attempts at resuscitation were unsuccessful.

Autopsy Findings

The autopsy was limited to the heart. The heart with attached aorta weighed 885 g. The pericardial sac was obliterated by fibrous adhesions. Blood obtained from the right heart had negative results on culture. A Faxitron radiograph of the heart specimen revealed a Björk-Shiley prosthesis in correct position. Sagittal section revealed no evidence of prosthetic valve dehiscence, thrombosis or infection. The aortic root was surrounded by scar tissue. Small linear scars on the aortic wall of the right sinus of Valsalva indicated adequate healing of the repaired aneurysm. The adjacent ventricular septal myocardium and the membranous septum were incorporated in a collagenized scar that occupied the entire width of the ventricular septum in the anteroposterior direction. Scar tissue was estimated to involve at least one-third of the entire ventricular septum in apical-basal direction. The endocardium covering the left ventricular outflow tract over the scar was markedly thickened (fig. 6). A rupture site was incorporated in the endocardial scar. The configuration of the entire heart was abnormal due to marked dilatation and hypertrophy of its chambers, particularly the left and right ventricle. The remainder of the aorta was normal, and there was no evidence of coarctation of the aorta or cystic medial necrosis. The coronary arteries also were entirely normal. Histologic examination of the ventricular septal scar revealed suture granulomas with an active chronic inflammation and well-collagenized scar. The non-
Sinus of Valsalva aneurysms are found in approximately 3.5% of patients with surgically treated congenital heart disease. These lesions are most commonly congenital, although Marfan's syndrome, endocarditis and, rarely, syphilis have been associated etiologically. The unruptured congenital sinus of Valsalva aneurysm has been reported to present with unexplained arrhythmias, right ventricular outflow obstruction or tricuspid insufficiency, aortic regurgitation, or coronary occlusion. The clinical significance and manifestations of ruptured sinus of Valsalva aneurysms depend on the direction and amount of blood flow through the abnormal intracardiac communication resulting from the rupture. Although site of rupture may vary widely, perforation most often occurs into the right cardiac chambers, resulting in acute left and right ventricular volume overload and commonly progressing to relentless congestive cardiac failure.

Although a defect in the membranous intraventricular septum is a commonly associated lesion, dissection of the sinus of Valsalva aneurysm into the interventricular septum is rare (table 2). All previous reports have been based on postmortem diagnoses. Examination of the data from these reports reveals a high incidence of interventricular conduction disturbances, but no other distinguishing clinical characteristics. The findings in the present case suggest that the diagnosis of sinus of Valsalva aneurysm with dissection into the muscular interventricular septum can be made preoperatively by means of echocardiography and angiocardiography.

Scarred myocardium exhibited hypertrophy, mild interstitial fibrosis and no evidence of myocardial infarction. The mitral valve was entirely normal.

FIGURE 5. Postoperative M-mode echocardiographic sweep from aorta to left ventricle. Dissection of the interventricular septum (arrowhead) is no longer apparent.

FIGURE 6. Sagittal cut through the heart, showing left ventricular inflow and outflow tracts. View of the cut surface of the posterior half of the heart. Arrow points to the deep scar that resulted from healing of the intraventricular septal defect. RV = right ventricle; LV = left ventricle.
Previous reports have revealed a wide assortment of echocardiographic abnormalities associated with sinus of Valsalva aneurysms. Aneurysms involving the right coronary sinus produce an abnormal echo anterior to the anterior aortic wall,14 occasionally with visualization of the defect in the aortic wall through which the right coronary cusp crosses in systole,15-20 as in the present case. Aneurysms of the left coronary sinus produce abnormal linear echoes posterior to the posterior aortic wall.14, 21 Echocardiographic findings other than visualization of the aneurysmal wall depend upon the presence and site of rupture and intracardiac communication. Rupture into the right heart is associated with systolic and diastolic fluttering of the anterior tricuspid leaflet19, 23 and premature pulmonic valve opening due to torrential flow resulting in rapid rise in diastolic right ventricular pressure to levels exceeding that in the pulmonary artery.24 Protrusion of an unruptured right coronary sinus of Valsalva aneurysm into the left ventricle adjacent to the intraventricular septum produces an abnormal group of echoes immediately posterior to the intraventricular septum, with characteristic abrupt early diastolic posterior motion.25-27

The present case showed most of the echocardiographic features of right coronary sinus of Valsalva aneurysm. The anterior aortic defect was demonstrated, as well as movement of the remnant of the right coronary cusp through the defect in systole (fig. 2), with prolapse into the left ventricular outflow tract during diastole (fig. 3). A unique finding was visualization of a longitudinal defect in the intraventricular septum continuous with the aneurysmal chamber (fig. 3), representing the unusual occurrence of septal dissection. Features that distinguished our patient’s condition from unruptured right coronary sinus of Valsalva aneurysm with protrusion into the left ventricle25-27 included absence of the typical diastolic motion pattern of the aneurysm wall described above and separation of septal echoes with reconstitution below the level of the mitral valve leaflets (fig. 1). The angiographic features of the present case were also distinctive, with a tract within the intraventricular septum that was opacified throughout the cardiac cycle (fig. 4).

The fact that all previously reported cases were derived from autopsy data indicates that this lesion is associated with a poor prognosis. Although ventricular scarring with resultant predisposition to life-threatening ventricular arrhythmias could have contributed to this patient’s death, the poor outcome was probably related to irreversible left ventricular damage that occurred before surgery. The ability of M-mode and cross-sectional echocardiography to accurately define the anatomical lesion in the present case before catheterization and surgery, however, is encouraging, and suggests that the management of similar cases could be optimized by the use of these noninvasive techniques.

Acknowledgment

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