Two-Dimensional Echocardiographic Recognition and Repair of Subvalvular Mitral Aneurysm of the Left Ventricle in an Infant

ROBERT SHADDY, MD, NORMAN H. SILVERMAN, MD, FACC, PAUL STANGER, MD, FACC, PAUL EBERT, MD, FACC

San Francisco, California

The clinical features, diagnostic studies and surgical treatment of a subvalvular mitral aneurysm of the left ventricle are described. The infant presented at 9 weeks of age with large apical ventricular septal defects and pulmonary hypertension. The subvalvular aneurysm was an incidental finding. Both lesions were treated surgically.

(J Am Coll Cardiol 1985;5:765-9)

The term "anular subvalvular left ventricular aneurysm" was first used in 1962 by Abrahams et al. (1), who reported 12 cases of left ventricular aneurysm which arose in the fibrous rings below either the aortic or the mitral valve and extended around the valves. There have been subsequent reports of these aneurysms, primarily in Nigerian and South African Bantu patients (2-4), although it has been reported in North American black (5) and white (6,7) patients as well. Until recently, diagnostic methods have consisted of history and physical examination, electrocardiogram, chest X-ray film, floroscopy, angiography and pathologic findings. We describe a 9 week old infant in whom echocardiography played a major role in diagnosing a submitral left ventricular aneurysm. Surgical treatment of the aneurysm and closure of multiple apical ventricular septal defects were undertaken.

Case Presentation

The patient, a white male infant who was the product of a 43 week gestation period, was noted to have a heart murmur in the newborn period and soon thereafter developed tachypnea and hepatomegaly. Cardiac catheterization at 2 weeks of age was interpreted to show a left ventricular to right atrial shunt, and further investigation including echocardiography suggested the presence of an atrioventricular canal defect. Digoxin and furosemide were given to aid the treatment of cardiac failure. He was referred to the University of California, San Francisco at 9 weeks of age.

Physical findings. The patient's parents reported his easy tiring and sweating with feedings. His weight gain was poor. Physical examination showed an infant weighing 11 pounds 9 ounces (4,953 g). Heart rate was 130 beats/min, respiration was 72/min and blood pressure was 120/70 mm Hg in the right arm. The liver was palpated 4 cm below the right costal margin. There was no cyanosis, digital clubbing or edema. Pulses were brisk and equal in all limbs. The first heart sound was normal. The second heart sound was narrowly split with a markedly accentuated pulmonary component. There was a grade 2/6 holosystolic murmur heard best at the lower left sternal border. There was a grade 2/6 mid-diastolic murmur at the apex. The clinical findings were those of a ventricular septal defect with pulmonary hypertension.

An electrocardiogram showed a QRS axis of -30°, left and right ventricular hypertrophy and nonspecific ST-T wave abnormalities. Right ventricular conduction delay was present. The chest X-ray film showed cardiomegaly and increased pulmonary vascular markings compatible with left to right shunt.

Echocardiographic findings. An echocardiogram showed biventricular and right atrial enlargement. The ventricular contraction was slightly depressed. There was a saccular polylobar structure within the right atrium, which was thought to represent an aneurysm of the atrioventricular septum (Fig. 1). The neck of this aneurysm arose close to the medial portion of the mitral valve and communicated only with the left ventricle. A venous contrast echocardiogram showed...
that the polylobar saccular structure lying within the right atrium was not in communication with the right atrium (Fig. 2). A large apical ventricular septal defect with a small second defect was imaged by two-dimensional echocardiography. The finding of these defects suggested that there were multiple apical ventricular defects. Venous contrast echocardiography showed a right to left ventricular shunt through an apical muscular ventricular septal defect (Fig. 3). The aneurysm only filled after the left ventricle became opacified. M-mode echocardiography revealed evidence of left ventricular volume overload and signs of pulmonary hypertension. The clinical impression was that of multiple apical ventricular septal defects with pulmonary hypertension and associated subvalvular mitral ventricular aneurysm prolapsing through the atrioventricular septum into the right atrium.

**Cardiac catheterization.** The procedure was performed within 24 hours of admission. The hemodynamic data showed systemic systolic pressures in the right ventricle and pulmonary artery, a pulmonary to systemic flow ratio of 3:1 and pulmonary vascular resistance of 4.8 units. A left ventricular angiogram showed an enlarged, smooth-walled posterior left ventricle of normal contractility with a normal competent mitral valve and a large apical ventricular septal defect. The aneurysm arose from the left ventricular inflow area adjacent to the mitral valve and contracted paradoxically to the rest of the ventricle when filled with contrast material (Fig. 4). There was no mitral insufficiency or evidence of an atrioventricular canal defect. A right atrial angiogram showed a large aneurysmal dilation of the left ventricular inflow area impinging extensively into the right atrium.

**Surgical treatment.** At surgery, an apical ventriculotomy was made in the right ventricle, and two apical ventricular septal defects were identified and closed with Teflon patches. A solid mass was palpable in the central portion of the right atrium, and this mass pulsated with ventricular systole. The right atrium was opened with a long oblique incision, and a large multilobed mass protruding into the right atrium was then opened longitudinally (Fig. 5). The wall was thick and fibrous and communicated with the left ventricle at the neck, which was approximately $1.5 \times 1$ cm. A counterincision was then made in the left atrium,
and the mitral orifice and mitral valve appeared normal. Interrupted sutures were placed around the orifice of this fibrous defect and a Teflon patch was tied in place. The aneurysm was thinned to reduce its bulk and then oversewn on the right atrial side. The patient had an uneventful postoperative recovery.

**Follow-up.** An echocardiogram done 3 days after operation showed a residual apical ventricular septal defect, with right to left shunting by contrast echocardiography.

**Figure 4.** Left ventricular (LV) angiogram showing filling of the aneurysm (An) from the left ventricle. The arrows indicate the multilobed character of the aneurysm. Ao = aorta.

**Figure 5.** Surgical view through the opened right atrium showing the bilobed aneurysm (An) located adjacent to the atrial septum and above the tricuspid valve orifice (TV).
The right atrial mass had been effectively excised, and the atrioventricular septum appeared normal with the mitral valve anulus abutting the septum. The patient underwent repeat cardiac catheterization at 8 months of age to evaluate the signs of persistent shunting. The catheterization data showed systolic pressure in the right ventricle and pulmonary artery to be half the systemic pressure, a pulmonary to systemic flow ratio of 2:1 and pulmonary vascular resistance of 4 units. Data also suggested a residual apical ventricular septal defect with moderate left to right shunt, moderate pulmonary hypertension and mild mitral insufficiency. There was no residual left ventricular aneurysm.

Discussion

Submitral left ventricular aneurysm is an uncommon anomaly. Most reported cases are in black African patients, and there are very few reports from white patients in the United States. The aneurysm occurs just below the mitral valve and extends around and into the substance of the fibrous anulus of the valve (1). Commonly, the aneurysm is single, but they may be multiple also. It usually has multiple orifices, but in our patient there was only one orifice although the aneurysm was multilobular. Some aneurysms have been described as compressing the left atrium (1). However, in our case, the aneurysm eroded through the fibrous atrioventricular septum and occupied the right atrium. Compression of the left coronary artery may also occur, which can lead to myocardial ischemia or infarction (2).

Etiology. The etiology of these aneurysms is unknown. This type of aneurysm is clearly not related to coronary artery disease, malaria, bacterial endocarditis, syphilis, tuberculosis, myotic infection, trauma, Loeffer’s parietal endocarditis, anomalous origin of a coronary artery from the pulmonary trunk, polyarteritis, tumors or cysts (1,3,7). Abrams et al. (1) speculated that a congenital defect or weakness of the ventricular wall in the region of the atrioventricular groove resulted in formation of an aneurysm. The pressure of the left ventricle then causes a herniation of the endocardium in that region, forming one or more ostia connecting with the ventricle. The overwhelming black racial predominance of this lesion has also led to speculations of a genetic origin. To our knowledge, our patient at 9 weeks of age represents the youngest report of surgical repair of a submitral left ventricular aneurysm. His young age and absence of other etiologic factors strongly support the congenital nature of this lesion.

Hemodynamic consequences. The hemodynamic consequences of these aneurysms include mitral regurgitation as a result of 1) dilation or distortion of the anulus produced by the aneurysm, or 2) failure of leaflet apposition caused by ballooning of the posterior leaflet (4). In our patient, mild mitral insufficiency did not become evident until after surgery. The diastolic load on the left ventricle may be further increased by the blood from the aneurysm emptying into the left ventricle, resulting in hypertrophy and congestive heart failure. Thrombus formation and systemic emboli have also been reported as a result of left ventricular aneurysm (3,8).

Histology. Histologically, the aneurysms showed marked endocardial thickening with a layer of dense collagen superimposed on the elastic layer of the endocardium. Calcification is not uncommon, particularly in older patients (8).

Clinical findings. The clinical presentation varies considerably. Reasons for cardiac consultation include congestive heart failure (1), hemiplegia (1,3,9), chest pain (2,6), heart murmur (7) and abnormal cardiac contour or calcification in an asymptomatic patient (8). In some instances, the lesion has been undiagnosed, and indeed unsuspected, during life (1,4). There are no diagnostic auscultatory signs. Systolic and diastolic murmurs and diastolic gallop sounds are frequently present (6). The presence of a nonejection systolic click or late systolic murmur of mild mitral regurgitation has been described (4). In our patient, there were no signs or symptoms related to the aneurysm and the physical findings were those of a ventricular septal defect with pulmonary hypertension.

Electrocardiographic findings. The electrocardiogram usually shows nonspecific ST-T wave changes, often with signs of myocardial ischemia or infarction. Other abnormalities include left ventricular hypertrophy, low voltage, conduction disturbances and arrhythmias (4).

Radiographic findings. Cockshott et al. (8), reviewed the radiologic features of 46 patients with anular subvalvular left ventricular aneurysm. The cardiothoracic ratio was increased in all but one case. The shape of the heart was characteristic in more than two-thirds of the patients due to a bulge that was present somewhere on the left heart border. Myocardial calcification was detected in approximately one-quarter of the patients and was more frequent in those without symptoms at the time of presentation. Oblique views or a penetrated anterior view often helped delineate the aneurysm. However, it was evident from angiography that intracardiac extension of an aneurysm could so displace cardiac chambers that from conventional frontal, lateral and oblique views it was frequently impossible to detect the aneurysm. Such was the case in our patient, whose only abnormalities were cardiomegaly with increased pulmonary vascular markings. Fluoroscopy may demonstrate systolic expansion of the bulge on the left cardiac border, although this was demonstrated in fewer than one-quarter of the patients described by Cockshott et al. (8).

Echocardiographic findings. M-mode echocardiography may suggest the diagnosis (10), but two-dimensional echocardiography offers the potential to make this diagnosis noninvasively while the patient is alive and to find associated defects. Detection of the aneurysm would depend on what
position it occupies. The erosion through to the right side of the heart is unusual. Under such circumstances, contrast echocardiography may be extremely helpful in outlining the aneurysm in silhouette, as it was in this case.

Before the development of two-dimensional techniques such as echocardiography, computed tomography and nuclear magnetic resonance imaging, the procedure of choice in investigating these aneurysms had been left ventricular angiography. With a left ventricular injection, the aneurysm is delineated and remains opacified after the ventricular cavity has emptied of contrast material. Left ventricular angiography also demonstrates any incompetence of the mitral valve. If the coronary arteries cannot be visualized well on the left ventricular angiogram, an ascending aorta angiogram should be performed. Left or right atrial angiography, or both, may be useful in demonstrating the intracardiac extension and compression of the aneurysm.

Surgical treatment. Operative repair of the aneurysm seems to be the most appropriate management (7). The risks associated with medical management alone include congestive heart failure, thromboembolic phenomena, acute coronary insufficiency and the risk of rupture (11). When Abrahams et al. (1) presented their series in 1962, they believed that surgery offered the only rational treatment. However, cardiopulmonary bypass was not available at that time. With the recent advances made in cardiopulmonary bypass, particularly in small infants (10), the morbidity and mortality associated with the surgical repair of this lesion should be minimal. To our knowledge, this paper represents the youngest reported patient with this anomaly and the first time echocardiography was successfully used in diagnosing this lesion.

References