Left Ventricular Diverticulum and Mitral Incompetence in Asymptomatic Children

M. Gueron, M.D., M. Hirsch, M.D., I. Opschitzer, M.D., and P. Mogel, M.D.

Two children with congenital apical left ventricular diverticulum and significant mitral incompetence are reported. The angiographic and anatomic findings of the few previously reported patients with congenital diverticula and aneurysm were analyzed and a new classification differentiating between the two is proposed.

An aneurysm or aneurysmal dilation of the left ventricle may complicate anomalous origin of the left coronary artery from the pulmonary artery. However, true congenital aneurysm of the left ventricle in children is very rare. Only a few scattered reports are available and in these the aneurysm was attributed to cardiomyopathy or to a congenital muscular defect of the left ventricle.

In previous communications the terms aneurysm and diverticulum have been used interchangeably. Serious attempts were not made to differentiate between congenital left ventricular aneurysm and the different types of diverticula. However, from a review of the few previously reported cases, it is possible to classify the diverticula and to differentiate them from a true congenital aneurysm.

This report presents two asymptomatic children with left ventricular apical diverticulum of undetermined cause accompanied by severe mitral incompetence, a combination never previously observed in children. Anatomic and angio-graphic differences between aneurysm and diverticula are defined and the appropriateness of surgery in such patients is discussed.

Case 1

This two-year-old bedouin girl was hospitalized in February, 1974, for kerosene pneumonitis. According to this history she was the product of a normal pregnancy and delivery. Prior to present admission she was hospitalized three additional times for recurrent gastroenteritis and pneumonitis, at the age of four and ten months, respectively. (Unfortunately we do not have any information concerning her cardiovascular status during these three hospitalizations in 1972.)

On admission in 1974, the physical examination revealed a well-developed girl without cyanosis or signs of congestive failure. The systolic blood pressure was 90 mm Hg. The peripheral pulses were equal and normal on palpation. The heart size was enlarged with a widespread, lifting, cardiac impulse palpable at the 6th left intercostal space in the anterior axillary line. There were no thrills, but a definite left ventricular heave was present. The first heart sound was normal, the second sound at the second left intercostal space was single and did not split with respiration. A loud third
sound was heard at the apex simulating a diastolic gallop. A grade 3/6 blowing musical pansystolic murmur, maximal at the apex was heard; the murmur radiated toward the left axilla and to the left lower sternal border. The liver and the spleen were not enlarged.

The electrocardiogram (fig. 1) and the vectorcardiogram showed left ventricular hypertrophy and a pattern consistent with apical myocardial infarction. The chest roentgenogram showed cardiomegaly probably due to the left ventricular enlargement (fig. 2).

The clinical data were highly suggestive of an anomalous left coronary artery originating from the pulmonary artery and secondary mitral incompetence due to papillary muscle dysfunction.

On catheterization the resting hemodynamics of the left ventricle were normal. The left ventricular angiogram revealed an apical diverticulum connected through a relatively short and narrow channel to the left ventricular cavity; severe mitral incompetence was also present (fig. 3). There was also delayed emptying of the diverticulum. A supravalvular thoracic aortogram showed a normal right preponderant coronary artery; the left coronary artery was slightly hypoplastic with a diminutive left anterior descending branch (fig. 4). No vessels were identified on the wall of the diverticulum. The apical region was avascular.

The patient recovered from the kerosene pneumonitis and was lost to follow-up.

Case 2

This 18-month-old bedouin girl was hospitalized in October 1974 for evaluation of her cardiac status. Her mother observed that she had a prominent pulsating "heart" and requested hospitalization. No previous history was available.

The physical examination revealed a well-developed child without heart failure. The blood pressure was 110/60 mm Hg. The heart was enlarged with a lifting widespread cardiac impulse palpable at the 6th left intercostal space, 1 cm before the anterior axillary line. There were no thrills, but a definite left ventricular heave was present. The first heart sound was normal, the second sound at the second left intercostal space was also normal and physiologically split. A grade 3/6 harsh musical pansystolic murmur maximal at the apex was heard radiating toward the left axilla and along the left sternal border, losing the harsh quality and becoming blowing. The phonocardiogram also recorded a fourth heart sound. The spleen and the liver were not palpable.

The electrocardiogram (fig. 5) and the vectorcardiogram showed left ventricular hypertrophy and changes consistent with lateral myocardial infarction. The chest roentgenogram
showed cardiomegaly with enlargement of the left atrium (fig. 6).

The clinical findings were consistent with an anomalous origin of the left coronary artery with secondary mitral incompetence due to papillary muscle dysfunction.

The left ventriculogram (fig. 7) showed a localized outpouching of the left ventricular apex connected to the enlarged ventricular cavity through a long narrow channel. Extreme opacification of the left atrium due to severe mitral incompetence was also observed. A supravalvular thoracic aortogram revealed normal coronary arteries with a prominent avascular apical area (fig. 8).

The patient was followed in the cardiac clinic and she remained asymptomatic. The electrocardiogram and the chest roentgenogram remained unchanged.

Discussion

Left ventricular aneurysm is classified as congenital when no known causes of acquired aneurysm are discovered. In the past the terms congenital aneurysm and diverticulum have been used interchangeably. Adequate attempts were not made to differentiate between the two, and the differences published were not distinct. It is evidently not only a problem of definition, but also a problem of understanding the basic pathology of the two abnormalities.

When the connection to the left ventricular cavity was narrow and there were thoraco-abdominal defects, the lesion was labeled diverticulum; the term aneurysm of left ventricle was applied when the opening at the point of connection was wide and there were no midline defects. This classification is not satisfactory since there are diverticula without thoraco-abdominal defects.

Chesler proposed the following classification based on the anatomical characteristics of the diverticulum (aneurysm?) of the left ventricle, a classification that did not include true apical aneurysmal dilation of the left ventricle as described in reports by Paronetto and Strauss and Ruttenburg et al.

Congenital muscular diverticulum of the heart is an outpouching including the myocardium, the endocardium, and occasionally the pericardium. The diverticulum may arise in the right, left, or both ventricles and is frequently reported in association with the intracardiac congenital defects presenting with cyanosis and midline defects. The muscular diverticulum typically arises from the apex of the ventricle and the point of connection to the left ventricular cavity is narrow and usually diagnosed early in life.

The fibrous diverticula of the left ventricle are located at the base most frequently but occasionally at the apex. The basal diverticulum is associated with the mitral and aortic rings, producing mitral or aortic incompetence. These subannular valvular diverticula may be multiple; the point of connection to the left ventricle is usually narrow at the level of the mitral valve annulus. Deposits of calcium, frequently reported in the fibrous type, have never been documented in the muscular type. Most of the patients are Negroes or Africans. The fibrous type is never accompanied by midline defects or intracardiac congenital heart malformations.

It is quite difficult to differentiate between true aneurysm and diverticulum on the basis of the histopathological findings since fibrosis and diminished myocardial fibers may be...
found in both. The presence of thoraco-abdominal deformities would suggest that the abnormality is a left ventricular muscular diverticulum.

The angiographic features and gross anatomic findings of previously published cases reveal another basis for differentiating between congenital aneurysm and congenital diverticulum of the left ventricle (table 1).

**Group A** consists of patients with an aneurysm of the left ventricle connected to the ventricular cavity by a wide opening, similar to the reported patients by Ruttenberg et al. and Paronetto et al.

The true congenital aneurysms in the two reported cases probably are the result of a congenital muscular defect. Their angiographic and roentgenographic features appear similar to those reported in patients with an anomalous origin of the left coronary artery from the pulmonary artery and in adults with acquired coronary artery disease. The congenital left ventricular aneurysm is located at the apex, with a fibrous wall and a wide ventricular connection, and is not accompanied by any other defects.

**Group B** is composed of patients with the different types of diverticula and may be subdivided into three subgroups:

**Subgroup 1** comprises patients with the muscular apical diverticula and thoraco-abdominal defects.

**Subgroup 2** includes patients with the fibrous type of diverticula.

**Subgroup 3** is formed by our two patients with an apical diverticulum and mitral incompetence and the three patients with a calcified apical diverticulum.

The pathology of the latter three patients is not known. However, since calcification is common in the second subgroup, it is quite possible that these three belong to the group of patients with fibrous diverticula. We classified them with our two patients with apical diverticulum and mitral regurgitation as a subgroup of unknown etiology.
Our two patients differ from all the patients in the above-mentioned classification. From an angiographic point of view the patients belong to the group of apical fibrous diverticula (table 1); however, mitral incompetence, which was not reported in any of the previously reported patients, was found in both patients. Mitral regurgitation is common in those with basal subannular mitral diverticula, but the cause of mitral incompetence is not clear in our two patients. We believe the papillary muscle involvement of the diverticula may explain this clinical finding.

A review of the literature revealed two patients who fulfill the criteria of true congenital left ventricular aneurysm. Seven additional patients could be classified as belonging to the group of patients with isolated apical diverticula, although in one the connection to left ventricular cavity was by a wide short stem.

In reviewing all previously reported patients with aneurysm and diverticulum of the left ventricle (table 2), females predominate, and all but one were asymptomatic at first examination. The defect was accidentally discovered in six by chest radiography. This could be related to the fact that the narrow point of connections between the diverticulum and the left ventricular cavity restricts the amount of regurgitant flow into the aneurysmal sac.

The ECG in all patients but one showed abnormal findings consistent with an anterolateral myocardial infarction and ST-T wave changes suggestive of a ventricular aneurysm, a feature reported in adult patients with ventricular aneurysms related to coronary artery disease. Furthermore, the two patients classified here as having true congenital aneurysms displayed similar ECG abnormalities. This suggests that the ECG alone, showing the above-mentioned changes, should not be considered the definitive tool for diagnosing anomalous origin of left coronary artery from the pulmonary artery. The ECG abnormalities are only diagnostic of myocardial damage or aneurysmal formation.

A clinical diagnosis of left ventricular diverticulum can be made and should be considered in the differential diagnosis of patients displaying the above-mentioned ECG changes with or without an abnormal chest roentgenogram. The presence or absence of mitral incompetence does not assure the correct diagnosis since this condition is observed in patients with papillary muscle dysfunction related to anomalous origins of left coronary artery as well as in patients with left ventricular diverticula. The left ventriculogram will differentiate between aneurysm formation and diverticula and will locate the defect anatomically.

It has been pointed out that most of the patients were

Table 2. Reported Patients with Aneurysm or Diverticulum of Left Ventricle

<table>
<thead>
<tr>
<th>Author</th>
<th>Age (yr)</th>
<th>Sex</th>
<th>MI</th>
<th>ECG of MI</th>
<th>Ap</th>
<th>Calc</th>
<th>Pathol</th>
<th>Aneurysm vs diverticulum</th>
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</thead>
<tbody>
<tr>
<td>Edwards⁴</td>
<td>8/12</td>
<td>F</td>
<td>No</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>No</td>
<td>Fibrous</td>
</tr>
<tr>
<td>Paronetto⁵</td>
<td>8/12</td>
<td>F</td>
<td>No</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>No</td>
<td>Fibrous</td>
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<tr>
<td>Papillon¹⁷</td>
<td>7</td>
<td>M</td>
<td>?</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>?</td>
</tr>
<tr>
<td>Dimich¹⁵</td>
<td>7</td>
<td>M</td>
<td>No</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>?</td>
</tr>
<tr>
<td>Shabetail¹⁶</td>
<td>12</td>
<td>M</td>
<td>No</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>?</td>
<td>Diverticulum</td>
</tr>
<tr>
<td>Cooley⁶</td>
<td>8</td>
<td>F</td>
<td>No</td>
<td>+</td>
<td>+</td>
<td>No</td>
<td>Fibrous</td>
<td>Diverticulum</td>
</tr>
<tr>
<td>Gueron</td>
<td>2</td>
<td>F</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>No</td>
<td>?</td>
<td>Diverticulum</td>
</tr>
<tr>
<td>Gueron</td>
<td>1 ½</td>
<td>F</td>
<td>+</td>
<td>+</td>
<td>+</td>
<td>No</td>
<td>?</td>
<td>Diverticulum</td>
</tr>
<tr>
<td>Dubbi¹⁸</td>
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<td>F</td>
<td>No</td>
<td>+</td>
<td>+</td>
<td>No</td>
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<td>Diverticulum</td>
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<tr>
<td>Cheeler²</td>
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<td>+</td>
<td>+</td>
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<tr>
<td>Drennan¹⁹</td>
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<td>?</td>
<td>?</td>
<td>+</td>
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<tr>
<td>Vivas-Salas²⁰</td>
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<td>M</td>
<td>?</td>
<td>RVH</td>
<td>+</td>
<td>No</td>
<td>?</td>
<td>Diverticulum</td>
</tr>
</tbody>
</table>

Abbreviations: Ap = apical location; Calc = presence of calcifications; Pathol = pathological classifications; RVH = right ventricular hypertrophy; MI = mitral incompetence.
asymptomatic at the time of presentation. The question arises, is surgical treatment justified since the natural history is not known? Review of the patients does not give a definite answer: two patients by were operated on, one for unstated reasons and the second because of the possibility of rupture and systemic embolism.

The problem becomes more complicated when mitral incompetence is present; despite successful surgical correction of one adult patient with a subannular mitral diverticulum, it is evident that surgical correction may be difficult. Furthermore, the suggestion that the diverticulum be excised when there is evidence of increasing size or severe mitral incompetence has yet to be supported with clinical evidence.

In conclusion, classification of previously reported cases makes it possible to differentiate between true congenital aneurysm and congenital diverticulum of the left ventricle. The differentiation is based mainly on angiographic as well as on pathological findings. As seen in our patients, the presence of mitral incompetence does not automatically indicate a subannular mitral diverticulum, and valvular incompetence may be encountered in apical diverticula.

The natural course of patients with apical diverticula is not known in asymptomatic children. Although mitral incompetence may be an additional burden on left ventricular function in these patients with apical diverticula, the place of surgery in the treatment of children or infants with this defect is not yet clear.

References
Left ventricular diverticulum and mitral incompetence in asymptomatic children.
M Gueron, M Hirsch, I Opschitzer and P Mogel

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