Congenital Absence of the Left Pericardium

Clinical, Electrocardiographic, Radiographic, Hemodynamic, and Angiographic Findings in Six Cases

By William K. Nasser, M.D., Charles Helmen, M.D., Morton E. Tavel, M.D., Harvey Feigenbaum, M.D., and Charles Fisch, M.D.

SUMMARY

Until the past decade, the diagnosis of congenital absence of the pericardium, partial or complete, had rarely been made prior to postmortem examination or thoracotomy.

Since 1963, the condition has been recognized during life in six patients at this institution. Of these six patients, two had partial absence of the left pericardium and four had complete absence of the left pericardium. Characteristic roentgenologic findings were present in all six patients. Associated heart lesions were not present in either patient with a partial pericardial defect. Two of the remaining four had associated heart lesions. One patient had surgical repair of an atrial septal defect. Surgical repair of the pericardial defect was not attempted in any of the six patients. Hemodynamic determinations at rest were normal in all six patients. The two patients with partial pericardial defects, however, had elevation of the pulmonary artery and left ventricular end-diastolic pressures during mild exercise in the recumbent position which suggests that this type of defect is not totally innocuous. In view of the unusual and extreme cardiac mobility in this condition, it is conceivable that a portion of the heart could herniate and transiently incarcerate through the partial defect during exercise. It is suggested that partial pericardial defects may warrant surgical repair. Small defects or complete absence of the left pericardium, however, are apparently without lethal potential and do not require surgical intervention.

Additional Indexing Words:
Atrial septal defect Murmurs Cardiac herniation

UNTIL the past decade, the diagnosis of congenital absence of the pericardium, partial or complete, had rarely been made prior to postmortem examination or thoracotomy. Although M. Realdo Columbus¹ is credited with the first reported case in 1559, the first unquestionable example of this condition was reported by Baille² in 1793. In ensuing years, an occasional case was reported as an incidental finding at surgery or necropsy.³-⁵ It was not until 1959, however, that Ellis and associates⁶ reported the first case recognized roentgenologically. Subsequently, increasing numbers of cases have been reported. Since 1963, six cases have been recognized during life at this institution.

The purpose of this report is fivefold: (1) to confirm previous reports that this entity can be diagnosed during life; (2) to suggest that it is not as rare as once believed; (3) to familiarize both clinician and radiologist as to the existence of this condition; (4) to support previous contentions that absence of the pericardium, per se, does not significantly

From the Department of Medicine and the Department of Radiology, Indiana University School of Medicine, and the Krannert Institute of Cardiology, Marion County General Hospital, Indianapolis, Indiana.

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Table 1

Clinical Findings in Six Patients with Absence of Pericardium

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age (yr); sex</th>
<th>Presenting complaint</th>
<th>Presumptive diagnosis</th>
<th>Chest pain</th>
<th>Dyspnea</th>
<th>Systolic ejection murmur (grade)</th>
<th>Diastolic murmur</th>
<th>Second sound</th>
<th>ECG</th>
<th>Associated defects</th>
<th>Heart lesion</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>19 M</td>
<td>Abnormal chest x-rays</td>
<td>Idiopathic dilatation of PA</td>
<td>0</td>
<td>0</td>
<td>II/VI</td>
<td>0</td>
<td>Normal</td>
<td>Normal</td>
<td>Absence of first left rib; pectus excavatum</td>
<td>0</td>
</tr>
<tr>
<td>2</td>
<td>50 F</td>
<td>Chest pain</td>
<td>Left hilar adenopathy</td>
<td>+</td>
<td>+</td>
<td>I/VI</td>
<td>0</td>
<td>Normal</td>
<td>Normal</td>
<td></td>
<td>0</td>
</tr>
<tr>
<td>3</td>
<td>11 M</td>
<td>Chest pain</td>
<td>Absence of left pericardium</td>
<td>+</td>
<td>+</td>
<td>I/VI</td>
<td>III/VI</td>
<td>Loud P₂</td>
<td>RAD; RVH</td>
<td>Pectus excavatum; Eisenmenger's syndrome (ASD)</td>
<td>0</td>
</tr>
<tr>
<td>4</td>
<td>12 F</td>
<td>Murmur</td>
<td>PS</td>
<td>0</td>
<td>0</td>
<td>III/VI</td>
<td>0</td>
<td>Normal</td>
<td>RAD; IRBBB; CWR</td>
<td>Pectus excavatum</td>
<td>0</td>
</tr>
<tr>
<td>5</td>
<td>53 M</td>
<td>Murmur; dyspnea</td>
<td>ASD</td>
<td>0</td>
<td>+</td>
<td>III/VI</td>
<td>0</td>
<td>Fixed split</td>
<td>RAD; IRBBB; CWR</td>
<td>Pectus excavatum; ASD with APVR</td>
<td>0</td>
</tr>
<tr>
<td>6*</td>
<td>20 M</td>
<td>Abnormal chest x-rays</td>
<td>Absence of left pericardium</td>
<td>+</td>
<td>+</td>
<td>I/VI</td>
<td>0</td>
<td>Normal</td>
<td>IRBBB; CWR</td>
<td>Pectus excavatum</td>
<td>0</td>
</tr>
</tbody>
</table>

*Case has been previously reported.

Abbreviations: PS = pulmonary stenosis; 0 = not present; ASD = atrial septal defect; APVR = anomalous pulmonary venous return; CWR = clockwise rotation; RAD = right axis deviation; IRBBB = incomplete right bundle-branch block; + = present.
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alter cardiac function; and (5) to show hemodynamic data suggesting that partial deficiency of the left pericardium may not be totally innocuous.

Results

The six patients ranged in age from 11 to 53 years (table 1). Four of the six patients were male; all were Caucasian. Of the six patients, two had partial absence of the left pericardium, and four had complete absence of the left pericardium. Associated heart lesions were not present in either patient with a partial pericardial defect. Two of the other four had associated lesions; one patient (case 5) had an atrial septal defect with partial anomalous venous drainage to the superior vena cava which was repaired surgically. Complete absence of the left pericardium was confirmed at operation, but no attempt was made to repair the pericardial defect. The patient's postoperative course was uneventful. The other patient with an associated cardiac lesion (case 3) had pulmonary hypertension with a reversed interatrial shunt (Eisenmenger's syndrome). Surgery was not performed on this patient nor on the other four patients. Chest pain was present in three patients and dyspnea in four. All six patients had systolic ejection murmurs of grade I to III on the basis of VI, which were heard best at the pulmonic area and left sternal border. Both patients with partial defects had normal electrocardiograms. The other four patients manifested electrocardiographic changes which consisted of right axis deviation (three patients), incomplete right bundle-branch block pattern (three patients), clockwise rotation in the horizontal plane (three patients), and right ventricular hypertrophy (one patient). Five of the six patients had an associated pectus excavatum deformity of the chest confirmed by physical examination and chest roentgenogram.

Cardiac catheterization, right and left, was performed on all six patients (table 2). The presence of intracardiac shunts was confirmed in two patients (cases 3 and 5) by oximetry, indicator-dilution techniques, and selective cinecardioangiography. The other four patients all had normal resting intracardiac pressures and no hemodynamic evidence of an associated cardiac lesion. Both patients with partial absence of the pericardium (cases 1 and 2) had elevation of the pulmonary artery and left ventricular end-diastolic pressures during mild exercise, which consisted of raising and lowering the left lower extremity for 4 minutes while in the recumbent position. The cardiac output and cardiac index were normal during rest and increased appreciably with exercise in three of the four patients with no associated cardiac lesion (cases 1, 2, and 6). The pulmonary vascular resistance was also normal in these three patients. The fourth patient without any associated cardiac lesion (case 4) was not exercised.

Roentgenographic studies revealed prominence of the pulmonary artery in all six patients (table 3, fig. 1). The left atrial appendage was prominent only in the two patients with partial defects. In addition to prominence of the pulmonary artery, all four patients with complete left pericardial defects had characteristic roentgenograms, namely, levoposition of the heart without tracheal deviation and interposition of a segment of lung between the aorta and pulmonary artery and between the inferior border of the heart and left hemidiaphragm (fig. 2A and B). These characteristic x-ray findings were not present in the two patients with partial or foramen-type defects. Artificial left pneumothorax (fig. 3) was diagnostic in the four patients on whom it was performed (cases 1, 3, 5, and 6). Herniation of the left atrial appendage through a pericardial foramen, as visualized by cineangiography, documented case 2 (fig. 4), and the other patient (case 4) had the characteristic roentgenographic findings of complete absence of the left pericardium (fig. 5). Cineangiography was performed in two patients with complete left pericardial defects but was not diagnostic. Results of pulmonary function tests performed on three patients (cases 2, 3, and 6) were normal.

Discussion

The rarity of this anomaly in the past is indicated by the fact that only two instances
### Table 2

**Hemodynamic Findings in Six Patients with Absence of Pericardium**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Condition</th>
<th>RA mean (mm Hg)</th>
<th>RV (mm Hg)</th>
<th>PA mean (mm Hg)</th>
<th>PCW mean (mm Hg)</th>
<th>LV (mm Hg)</th>
<th>CAo (mm Hg)</th>
<th>(Fick) CO (L/min)</th>
<th>(Fick) CI (L/min/m²)</th>
<th>PVR (dynes sec cm⁻²)</th>
<th>Type of pericardial defect</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Rest</td>
<td>5</td>
<td>26/7</td>
<td>26/9</td>
<td>14</td>
<td>8</td>
<td>128/9</td>
<td>128/80</td>
<td>5.5</td>
<td>2.9</td>
<td>80</td>
</tr>
<tr>
<td></td>
<td>Exercise</td>
<td>40/8</td>
<td>40/15</td>
<td>24</td>
<td>164/19</td>
<td>11.2</td>
<td>164/82</td>
<td>134/66</td>
<td>5.4</td>
<td>3.5</td>
<td>88</td>
</tr>
<tr>
<td>2</td>
<td>Rest</td>
<td>3</td>
<td>26/3</td>
<td>26/9</td>
<td>15</td>
<td>9</td>
<td>131/12</td>
<td>156/86</td>
<td>10.8</td>
<td>7.0</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Exercise</td>
<td>39/19</td>
<td>33</td>
<td>150/20</td>
<td>112/6</td>
<td>115/80</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>Rest</td>
<td>1</td>
<td>90/4</td>
<td>90/44</td>
<td>56</td>
<td>115/6</td>
<td>115/73</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Exercise</td>
<td>21/4</td>
<td>21/6</td>
<td>13</td>
<td>115/6</td>
<td>115/73</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>Rest</td>
<td>1</td>
<td>15/0</td>
<td>15/6</td>
<td>11</td>
<td>3</td>
<td>93/10</td>
<td>93/50</td>
<td>5.3</td>
<td>2.9</td>
<td>120</td>
</tr>
<tr>
<td></td>
<td>Exercise</td>
<td>20/1</td>
<td>20/3</td>
<td>10</td>
<td>108/12</td>
<td>108/55</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Abbreviations: RA = right atrium; RV = right ventricle; PA = pulmonary artery; PCW = pulmonary capillary wedge; LV = left ventricle; CO = cardiac output; CI = cardiac index; PVR = pulmonary vascular resistance; CAo = central sorta.

### Table 3

**Roentgenographic and Angiographic Findings in Six Patients with Absence of Pericardium**

<table>
<thead>
<tr>
<th>Patient</th>
<th>Prominence of PA</th>
<th>Levo-position</th>
<th>Interposition of lung between PA and sorts</th>
<th>Heart and diaphragm</th>
<th>Diagnostic pneumothorax</th>
<th>Cine-angiogram</th>
<th>Pulmonary function tests</th>
<th>Type of pericardial defect</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>+</td>
<td>+</td>
<td>0</td>
<td>0</td>
<td>Positive</td>
<td>Positive</td>
<td>Not done</td>
<td>Partial</td>
</tr>
<tr>
<td>2</td>
<td>+</td>
<td>+</td>
<td>0</td>
<td>0</td>
<td>Not done</td>
<td>Positive</td>
<td>Normal</td>
<td>Partial</td>
</tr>
<tr>
<td>3</td>
<td>+</td>
<td>0</td>
<td>+</td>
<td>+</td>
<td>Positive</td>
<td>Not diagnostic</td>
<td>Normal</td>
<td>Complete</td>
</tr>
<tr>
<td>4</td>
<td>+</td>
<td>0</td>
<td>+</td>
<td>+</td>
<td>Not done</td>
<td>Not diagnostic</td>
<td>Not done</td>
<td>Complete</td>
</tr>
<tr>
<td>5</td>
<td>+</td>
<td>0</td>
<td>+</td>
<td>+</td>
<td>Positive</td>
<td>Not diagnostic</td>
<td>Not done</td>
<td>Complete</td>
</tr>
<tr>
<td>6</td>
<td>+</td>
<td>0</td>
<td>+</td>
<td>+</td>
<td>Positive</td>
<td>Not done</td>
<td>Normal</td>
<td>Complete</td>
</tr>
</tbody>
</table>

Abbreviations: PA = pulmonary artery; LAA = left atrial appendage; + = present; 0 = not present.
were encountered by Versé\textsuperscript{5} in 13,000 autopsies and only one by Southworth and Stevenson\textsuperscript{8} in 14,000 autopsies at Johns Hopkins Hospital. In 1937, Dahl\textsuperscript{4} inferred the presence of a communication between the pericardium and left pleural space following artificial pneumothorax which resulted in pneumopericardium in a young man being treated for bilateral exudative pulmonary tuberculosis. It was not until 1959, however, that Ellis and co-workers\textsuperscript{6} described the characteristic roentgenologic features of plain chest roentgenograms and diagnostic pneumothorax. In 1960, Dimond and associates\textsuperscript{9} diagnosed the first case of a partial defect by cineangiography, demonstrating the herniated left atrial ap-
Figure 2

Postero-anterior (A) and left anterior oblique (B) views in case 6. In A the heart is shifted to the left (levoposition), the right heart border is hidden by the spine, the trachea is in the midline, the pulmonary artery segment is prominent (arrow), and there is interposition of lung between the left hemidiaphragm and inferior border of the heart. In B the arrow points to a tongue of lung separating the aorta and main pulmonary artery. Reprinted courtesy of Circulation from Nasser W, et al: Congenital Absence of the Left Pericardium. Circulation 34:100-104, 1966.

Figure 3

Diagnostic pneumothorax following the injection of 500 cc of air into the left thorax in case 1. In the left lateral decubitus view of the chest, pneumopericardium is produced and the right pleuropericardium (arrows) is outlined against air in the right lung.

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the pericardium has a male-to-female ratio of approximately 3 to 1.

Unless there is an associated cardiac anomaly, most of the reported patients with pericardial defects have been asymptomatic, however, symptoms are not infrequent. The most common symptom is usually vague chest pain. It has been suggested that the pain could result from torsion of the great vessels due to absence of the stabilizing forces of the left pericardium. Dyspnea, dizziness, and syncope have occasionally been present.

Physical findings are usually not helpful. Systolic ejection murmurs, noted in all six patients in the present series, are usually heard at the second left interspace or along the left sternal border. They may be the result of turbulence set up by varying mechanical deformities at the base of an unusually mobile heart. Precordial activity may be conspicuous and the apical impulse may be shifted to the left, especially in pericardial deficiencies of the entire left side. Aside from these few observations, the physical findings are dependent on the underlying cardiac disorder.

The electrocardiographic changes usually reflect the unusual intrathoracic position of the heart which is permitted by the absence of a restraining pericardium. Electrocardiographic abnormalities are more commonly seen in complete left pericardial defects and usually consist of right axis deviation, incomplete right bundle-branch block pattern, and leftward displacement of the transition in the precordial leads. Sinus bradycardia is not an uncommon finding. Complete heart block, presumably congenital, was reported in a single patient.

Characteristic roentgenologic findings are usually present in complete absence of the left pericardium (fig. 2). In partial pericardial defects, the heart is in normal position and the abnormality consists of varying degrees of prominence of the pulmonary artery or left atrial appendage or both (fig. 1). Cineangiography, demonstrating herniation of the left
atrial appendage beyond the left heart border, is considered diagnostic of partial absence of the left pericardium by several authors.30, 33-37 This procedure is usually not diagnostic for complete left-sided pericardial defects. Pneumopericardium, following induction of artificial pneumothorax, is the procedure of choice for diagnosing complete absence of the left pericardium. Since the x-ray findings are so strikingly similar, some authors18, 19, 35 consider that diagnostic pneumothorax, with its possible hazards, may no longer be necessary in establishing the diagnosis. Also, the absence of pneumopericardium following artificial pneumothorax does not rule out absence of the pericardium since, in some cases, adhesions could conceivably prevent the accumulation of air in the pericardial space,6, 13, 17, 19, 35

Experimental studies in which total or partial resection of the pericardium was performed in 65 dogs led to the conclusion that no impairment of cardiac function nor dilatation of the heart resulted in any of the animals.29 That disturbance of normal cardiac function does not appear to be present in patients with complete pericardial defects is suggested by two patients in the present series: both had large defects without intrinsic heart disease, and both had normal intracardiac hemodynamics (table 2, cases 4 and 6). Although both patients with partial defects had normal resting intracardiac pressure determinations (table 2, cases 1 and 2), the pulmonary arterial and left ventricular end-diastolic pressures did become abnormal with mild exercise in the recumbent position. Since there have been three reports of sudden death from herniation and strangulation of the heart through a partial pericardial defect,33, 38, 39 it would certainly appear that this type of defect is not totally innocuous. In view of the unusual and extreme cardiac mobility in this condition, it is conceivable that a portion of the heart could herniate and transiently incarcerate through the partial defect during exercise, resulting in elevation of the pulmonary artery and left ventricular end-diastolic pressures. This postulate could also explain the symptoms of chest pain, dyspnea, dizziness, and syncope which occasionally occur in the absence of intrinsic disease of the heart or lungs. Further support for this contention is suggested by the fact that (1) the pulmonary artery and left ventricular end-diastolic pressures did not elevate to abnormal levels following exercise in the patient (case 6) with a "normal" heart and complete absence of left pericardium; (2) there have been no reports of sudden death in patients with complete left-sided pericardial defects; and (3) pressure determinations during exercise have been normal in patients with complete absence of the left pericardium.32, 40

Surgical intervention for partial pericardial defects has been undertaken in recent years to prevent herniation and strangulation of the left atrium through the defect. The presence of symptoms and the possibility of sudden death in patients with partial defects, accompanied by angiographic evidence of herniation of the left atrial appendage through the pericardial defect, argue strongly for surgical correction of this lesion.13, 17, 41

Surgical procedures include left atrial appendectomy, division of adhesions, pericardio-plasty, or extension of the defect to make it larger. Small defects or complete absence of the left pericardium, however, are apparently without any lethal potential and do not require surgical intervention.

Addendum

Case 4, a patient with complete absence of the left pericardium and no associated cardiac lesion, has had a repeat heart catheterization since acceptance of this report. Her intracardiac pressure determinations including pulmonary artery and left ventricular end-diastolic pressures were normal during both rest and exercise.

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Words—Units of Communication

When I took the first survey of my undertaking, I found our speech copious without order, and energetick without rules: wherever I turned my view, there was perplexity to be disentangled, and confusion to be regulated; choice was to be made out of boundless variety, without any established principle of selection; adulterations were to be detected, without a settled test of purity; and modes of expression to be rejected or received, without the suffrages of any writers of classical reputation or acknowledged authority.

Having therefore no assistance but from general grammar, I applied myself to the perusal of our writers; and noting whatever might be of use to ascertain or illustrate any word or phrase, accumulated in time the materials of a dictionary, which, by degrees, I reduced to method, establishing to myself, in the progress of the work, such rules as experience and analogy suggested to me; experience, which practice and observation were continually increasing; and analogy, which, though in some words obscure, was evident in others.—SAMUEL JOHNSON: Preface to the Dictionary (1755).
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WILLIAM K. NASSER, CHARLES HELMEN, MORTON E. TAVEL, HARVEY FEIGENBAUM and CHARLES FISCH

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