Congenital Absence of the Left Pericardium

By William Nasser, M.D., Harvey Feigenbaum, M.D., and Charles Helmen, M.D.

UNTIL RECENT YEARS, the diagnosis of absence of the pericardium, partial or complete, had rarely been made prior to post-mortem examination or thoracotomy. There are presently less than 120 cases reported in the literature. The rarity of this anomaly is indicated by the fact that only two instances were encountered by Versé in 13,000 autopsies and only one by Southworth and Stevenson in 14,000 autopsies at the John Hopkins Hospital.

Although M. Realdo Columbu is credited with the original report of this entity in 1559, most authors consider that the first unquestionable example of this condition was reported by Baillie in 1793. Dah, in 1937, inferred the presence of a communication between the pericardium and left pleural space following artificial left pneumothorax which resulted in pneumopericardium in a young man being treated for bilateral exudative pulmonary tuberculosis. However, it was not until 1939 that Ellis and associates reported the first case recognized roentgenographically. Since this time, an increasing number of cases have been reported in the literature.

The purpose of this paper is threefold: (1) to confirm previous reports that this entity can be diagnosed during life; (2) to familiarize both clinician and radiologist as to the existence of this condition; and (3) to support previous contentions that absence of the pericardium, in itself, does not alter cardiac function.

Case Report

Case 1

This 20-year-old white male was admitted to the Indiana University Medical Center for evaluation of an enlarged heart. Six months prior to admission the patient had an annual chest x-ray and was found to have cardiomegaly.

He complained only of vague chest pain and dyspnea. These symptoms were not related to exertion, changes of position, or meals. At the age of 17, he was rejected for military service because of an enlarged heart.

Physical examination revealed normal blood pressure and pulse. Positive physical findings included a grade I/VI systolic ejection murmur at the second left intercostal space, a visible, diffuse, active point of maximal impulse in the fifth intercostal space 3 cm to the left of the mid-clavicular line, and a persistently split second heart sound, although it did widen with inspiration. All auscultatory findings were confirmed by phonocardiography. No thrills, heaves, peri-

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This investigation was supported by the Herman C. Krannert Fund, the Indiana Heart Association, and in part by U. S. Public Health Training Grant, 5363, and the facilities provided by the Cardiovascular Clinical Research Center Grant H-6308 from the National Heart Institute, U. S. Public Health Service.

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Figure 1

Posteroanterior roentgenogram of the chest. There is levoposition of the heart. The heart does not obliterate the diaphragm shadow. The pulmonary artery segment (arrow) is separated from the aortic shadow.
cardial friction rubs, diastolic murmurs, or gallops were present. The remainder of the physical examination was within normal limits.

The electrocardiogram showed incomplete right bundle-branch block with marked clockwise rotation and a normal axis.

Frontal roentgenograms revealed marked left ventricular dilatation. The main pulmonary artery segment appeared prominent due to interposition of the lung between the pulmonary artery and aorta (fig. 1). Parts of the lung were also visible below and behind the heart on the oblique projection (fig. 2). The overall heart size was considered normal. Fluoroscopy demonstrated normal pulsations.

Since complete absence of the left pericardium was strongly suspected, left pneumothorax was performed to confirm the diagnosis. Five hundred cubic centimeters of air was introduced into the left side of the thorax, and films were obtained in a left lateral decubitus position with a horizontal x-ray beam. The air was seen to enter the pericardial sac and to outline the right pleuropericardium (fig. 3). No air entered the right pleural space.

The presence of incomplete right bundle-branch block, persistent splitting of the second heart sound, systolic ejection murmur, and prominent pulmonary artery led to the clinical suspicion of atrial septal defect. Right and left heart catheterization was performed. Normal cardiac hemo-

**Figure 2**

*Right anterior (A) and left anterior (B) oblique roentgenograms: (A) Arrows show the lung around the heart; and (B) the arrow points to the tongue of lung separating the aorta and main pulmonary artery.*

**Table 1**

<table>
<thead>
<tr>
<th>Catheterization Data</th>
<th>Rest</th>
<th>Exercise</th>
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<tbody>
<tr>
<td></td>
<td>Pressure, mm Hg</td>
<td>O₂ sat. (%)</td>
</tr>
<tr>
<td>Right atrium</td>
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<td>73</td>
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<tr>
<td>Right ventricle</td>
<td>15</td>
<td>20</td>
</tr>
<tr>
<td>Pulmonary artery</td>
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<td>20</td>
</tr>
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<td>Pulmonary wedge</td>
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<td>10</td>
</tr>
<tr>
<td>Left ventricle</td>
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<td>10</td>
</tr>
<tr>
<td>Brachial artery</td>
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<td>108</td>
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<tr>
<td>Cardiac index</td>
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<tr>
<td>Pulmonary vascular resistance</td>
<td>1.5 units (rest)</td>
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</tbody>
</table>

*Circulation, Volume XXXIV, July 1966*
dynamics were found with no evidence of obstruction to outflow of either ventricle and no evidence of intracardiac or extracardiac shunt by oximetry or indicator-dilution techniques (table 1). The cardiac output was normal at rest and with exercise. Pulmonary function tests were within normal limits. Surgical intervention was not felt to be indicated.

Discussion

In their excellent postmortem review in 1938, Southworth and Stevenson,2 stated that 76% of the congenital pericardial defects reported in the literature reviewed involved the entire left side of the heart. The remainder were partial or total defects. This ratio is repeatedly quoted in subsequent reports. However, a review of the literature suggests that with the aid of pulmonary angiography partial left-pericardial defect is being reported7-13 more frequently than complete left-sided defect. Right-sided lesions and complete absence of the pericardium are extremely rare.

Associated congenital anomalies such as patent ductus arteriosus,7-14 bronchogenic cysts,15, 16 tricuspid insufficiency,17 atrial septal defects,18, 19 left diaphragmatic hernia,20 and pulmonary sequestration21 occur in approximately 30% of the cases. The condition has a male-to-female ratio of approximately 3 : 1.

In most of the reported cases pericardial deficiencies have been asymptomatic, and the condition has been suspected because of a peculiar appearance of the chest x-ray. Occasionally a patient with a partial defect has experienced chest pain.9, 13 It has been suggested that the chest pain could result from torsion of the great vessels with increased stress on the anchoring structures of the cardiac base due to absence of the stabilizing forces of the left pericardium.6 Three instances of death resulting from herniation and strangulation of the heart through a pericardial defect have been reported.22-24 Presumably, cardiac strangulation would not occur in the presence of a large pericardial defect.6

Physical findings in patients with pericardial defects are usually of no aid in diagnosis. Unexplained bradycardia has occasionally been observed. Systolic murmurs are frequently heard and may be the result of turbulence set up by varying mechanical deformations at the base of an unusually mobile heart6 and are most often heard in the second left-intercostal space. Precordial movement may be conspicuous, especially with large pericardial defects which allow unrestricted cardiac mobility. Aside from these few observations, the physical findings are dependent on any underlying cardiac disorder.

Figure 3

(A) Left lateral decubitus view of the chest following diagnostic left pneumothorax; and (B) line drawing of A—air is seen in the pericardial space and the right pleuropericardium is outlined against the air in the right lung.
The electrocardiogram may show marked changes in the electrical axis of the QRS complexes due to distortion of the normal intrathoracic position of the heart. There is a tendency to right axis deviation which makes the clinician suspicious of underlying congenital heart disease, especially intracardiac shunts.

Several functions have been given to the pericardium. These include: (1) isolation of the heart from infections of the lung and pleural space; (2) prevention of overdistention of the heart during the ventricular filling period; (3) protection of the lungs from the trauma of the beating heart; (4) maintenance of stable intracardiac pressures; and (5) mooring of the heart in optimum functional position.

It appears unquestionable that the pericardium does indeed offer protection to the heart from infections of the lung and pleural space. Southworth and Stevenson\(^2\) found that 27% of the patients had evidence of pleuroparcarditis, often secondary to infections of the lung. Since the introduction and widespread use of antibiotics, reported cases of pericarditis in documented pericardial defects have been infrequent.\(^9,13\)

A considerable amount of investigative work has been directed toward the function of the pericardium in restricting distensibility of the cardiac chambers. Kuno\(^25\) found that removal of the pericardium in the heart-lung preparation was followed by cardiac dilatation, lower filling pressures, elevated arterial pressure, increased cardiac output, and increased ventricular work. Holt and associates\(^26\) from observations on dogs, concluded that removal of the pericardium in plethoric states results in a greatly distended heart with high transmural pressures throughout the cardiac cycle. They felt that the heart was protected from overdistention in ventricular diastole when the pericardium was intact. It has also been suggested that the pericardium may offer protection against pulmonary edema by limiting right ventricular filling when the left ventricle is dilated.\(^27\) On the other hand, Moore and Shumacker\(^17\) performed total or partial resection of the pericardium in 65 dogs and concluded that no impairment of cardiac function nor dilatation of the heart resulted in any of these animals. Grant\(^28\) also concluded that the pericardium played no part in preventing overdistention of the heart and that its absence was not a factor in producing cardiac enlargement.

Berglund and associates\(^27\) concluded that insufficiency of the mitral and tricuspid valves develops more readily in the absence of the pericardium when the heart is subjected to increased filling pressure. Regurgitation through the atroventricular valves in dilated hearts was reported in experiments with open pericardium.\(^25,29\) Moore and Shumacker\(^17\) reported a 30-year-old female with marked right ventricular and right atrial enlargement associated with tricuspid insufficiency. At surgery, the entire presenting surface of the heart was deficient of pericardium. The etiology of the tricuspid insufficiency was obscure to these authors but, if the observations in experiments with the heart-lung preparation are valid, it seems possible that absence of the pericardium could have resulted in over-distention of the right ventricle with regurgitation into the atrium.

Although the exact functions of the pericardium have not been completely proved, the comments just made suggest that the pericardium does make some contribution, especially in the presence of inherent pulmonary or myocardial disease. However, the normal hemodynamic findings reported in this paper indicate that in the absence of diseases of the lungs or heart muscle, the pericardium, per se, is probably not essential for adequate cardiac function. This conclusion is supported by other reports of patients with pericardial defects and normal right-heart catheterization findings.\(^6,9-11,18,19\)

Thus, it appears that from a hemodynamic point of view, congenital absence of the left pericardium, per se, is a benign disease and should not be mistaken for serious heart disease, as was done in our patient.

Surgical intervention for partial pericardial defects has been undertaken in recent years
to prevent herniation and strangulation of the left atrium through the defect. This includes left atrial appendectomy, division of adhesions, pericardioplasty, 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.

Summary

A case of complete absence of the left pericardium, suspected from plain chest x-rays and proved by diagnostic left pneumothorax, has been reported. The significance of the pericardium in relation to cardiac function is discussed. Because absence of the left pericardium is a benign and relatively asymptomatic anomaly that usually requires no treatment, it behooves the clinician and radiologist to become cognizant of its presence.

References

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CIRCULATION. 1966;34:100-104
doi: 10.1161/01.CIR.34.1.100

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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