Congenital Absence of the Left Pericardium and Complete Heart Block*

Report of a Case

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THE RECOGNITION AND DEFINITIVE DIAGnosis of absent pericardium has gained impetus within recent years. Although well described for many years as a curious anomaly found at post-mortem examination, it was not until 1959 that Ellis et al. reported the first case of congenital absence of the pericardium in which the diagnosis was successfully established by diagnostic pneumothorax during life.

This case of deficient left pericardium with concomitant complete heart block depicted with angiographic, radiologic, and hemodynamic studies is presented and the proper diagnostic techniques underscored. The differentiation of complete and partial deficiency of the pericardium is stressed in terms of diagnosis, prognosis and treatment.

CASE REPORT:

This 29-year-old white man was admitted to St. Vincent's Hospital on April 29, 1964 for medical evaluation of an abnormal chest x-ray film and electrocardiogram noted during a routine examination.

He was first informed of a heart murmur at the age of 16 and, two years later, failed to pass the Armed Forces physical examination, presumably because of this cardiac murmur. He stated that he was in good health and offered no symptoms. The past medical history was negative.

The blood pressure was 130/88 mm Hg, pulse was 60 per minute and regular, and temperature was normal. He was well developed, with no cyanosis or respiratory distress. Cervical veins were flat in the recumbent position. Lungs were clear. The cardiac apical impulse was palpated in the sixth intercostal space and anterior axillary line and produced a moderate lift. No thrill

was palpable. The rhythm was regular. A grade II/VI ejection systolic murmur was audible along the left sternal border with maximal intensity in the second and third intercostal space. Splitting of the second sound occurred on inspiration. There was slight accentuation of the pulmonic component. No organ was palpable on abdominal examination. Moderate clubbing of the fingers was present. Peripheral pulses were strong.

Laboratory studies showed a hemoglobin of 15.7 mg per cent; hematocrit of 46 per cent; and white blood cell count of 9,500/mm³. Differential count was normal. Urinalysis, blood urea nitrogen and fasting blood sugar were all within normal limits. Chest x-ray film showed normal pulmonary vascularity and an abnormal cardiac contour. The heart was displaced to the left hemithorax with an apparent prominence of the left ventricular contour (Fig. 1). The electrocardiogram showed atrioventricular dissociation with complete heart block (Fig. 2). A vectorcardiogram was also done (Fig. 3).

Right heart catheterization showed normal hemodynamics (Table 1). Selective angiography

Figure 1: Significant features include leftward displacement of the heart without tracheal deviation. In addition, three distinct convexities are noted along the left border of the heart. These are the aortic knob, pulmonary artery segment and the left ventricular contour.

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FIGURE 2: Conventional 12 lead electrocardiogram shows atrioventricular dissociation with complete heart block. The ventricular rate (junctional rhythm) is 48 per minute. The transitional QRS complex of the precordial leads occurs between V₃ and V₄.

was performed (Fig. 4). The tricuspid valve was displaced to the left of the vertebral spine. The right ventricle appeared large and occupied the anterior half of the cardiac shadow. The outflow tract of the right ventricle and pulmonary artery were normal. The left ventricle occupied the posterior portion of the cardiac silhouette in the AP projection. The aorta was normal in position and origin. The possibility of absent pericardium was suggested by the findings on chest x-ray film. Hence, the patient was studied, using the technique of pneumothorax, with 500 ml of air introduced into the left pleural cavity and a collection of air was noted between the right visceral and parietal pericardium (Fig. 5).

He was discharged asymptomatic and has been well.

DISCUSSION

Congenital pericardial defects are predominantly left-sided. They present as either complete deficiency of the left pericardium, or as a partial defect, usually overlying the left pulmonary artery segment, or left atrium. The left-sided predilection has been ascribed to incomplete development of the left pleuropericardial membrane, secondary to premature atrophy of the left duct of Cuvier.

Strict differentiation must be made between partial pericardial defect and the complete variety. In the latter, the pericardial and pleural space form a common cavity and the heart, displaced toward the left, assumes an abnormal anatomic position within the chest. In partial pericardial deficiency, the heart maintains its normal posi-
FIGURE 4A: The tricuspid valve is displaced to the left of the vertebral spine. The right ventricular cavity is also displaced to the left and extends to the outermost contour of the heart. Enlargement of the right ventricle is also noted.

In this case, complete absence of the left pericardium which causes cardiac rotation to the left and posteriorly (pseudo-levorotation).
pericardium was suggested by two significant findings on chest roentgenograms. The most striking feature related to an abnormal shift of the heart to the left, with a normal tracheal position. Secondly, three distinct convexities were noted along the left cardiac border, consisting of the aortic knob, elongated pulmonary artery segment and flattened left ventricular contour.

The diagnosis was confirmed, however, only after utilization of left pneumothorax. Injection of 500 ml of air into the left pleural space produced a distinct pneumopericardium along the left cardiac border. This was discernable as air separating the right parietal and visceral pericardium.

The position most favorable to the demonstration of pneumopericardium, according to Ellis et al., is the right posterior oblique of the chest roentgenograms. This can be performed with the patient lying on the left side, with a horizontally directed beam. Another technique that permits accurate diagnosis is the lateral roentgenogram with the patient supine. In this position, the heart

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**TABLE 1—DATA OF CARDIAC CATHETERIZATION**

<table>
<thead>
<tr>
<th></th>
<th>Rest</th>
<th>Exercise</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right atrium</td>
<td>5</td>
<td>5</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>28/5</td>
<td>31/5</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>26/9 (12)*</td>
<td>29/9 (17)*</td>
</tr>
<tr>
<td>Pulmonary wedge</td>
<td>(8)*</td>
<td>(10)*</td>
</tr>
</tbody>
</table>

*Mean Pressure

<table>
<thead>
<tr>
<th></th>
<th>Rest</th>
<th>Exercise</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pulmonary artery</td>
<td>12.58</td>
<td>12.12</td>
</tr>
<tr>
<td>Brachial artery</td>
<td>18.47</td>
<td>18.54</td>
</tr>
<tr>
<td>Arterial oxygen content in volumes per cent</td>
<td>94.3%</td>
<td></td>
</tr>
<tr>
<td>Systemic blood flow liters/min</td>
<td>4.27</td>
<td>5.15</td>
</tr>
<tr>
<td>Systemic blood flow L/min/M²</td>
<td>2.17</td>
<td>2.61</td>
</tr>
<tr>
<td>Ventricular rate</td>
<td>50</td>
<td>56</td>
</tr>
<tr>
<td>Stroke volume</td>
<td>85 ml.</td>
<td>95 ml.</td>
</tr>
<tr>
<td>Hydrogen uptake studies — normal</td>
<td></td>
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</tr>
<tr>
<td>Dye dilution studies — normal</td>
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</tbody>
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**FIGURE 5A:** Diagnostic pneumothorax: Patient lying in the left lateral decubitus position. Pneumopericardium outlines the right pericardium (arrow). The heart shows further displacement to the left in this position.
is displaced posteriorly against the spine if the left pericardium is deficient. Left pneumothorax is an indispensable method of study if the diagnosis of complete deficiency of the pericardium is to be substantiated.

Angiocardiography, on the other hand, delineated the abnormal positional change of the heart, but was of no value in promoting a definitive diagnosis of absent pericardium. This technique, however, may be conclusive in partial deficiency, particularly when the left atrial appendage herniates beyond the pericardial border.

Of added interest in this case was the association of complete heart block, presumably of congenital origin. Pericardial defects have been described in association with a variety of congenital anomalies, including congenital heart disease, bronchogenic cysts, and aberrant pulmonary lobe. To our knowledge, this kind of conduction disorder has not been previously described with pericardial deficiency.

Despite the presence of these combined lesions, our patient was completely asymptomatic. There was no history of Adams-Stokes attack or dyspnea. Most patients with complete absence of the pericardium have been asymptomatic. Several cases have been described with unexplained chest pain, and one in which the pain was related to changes in body position.

Recurrent chest pain indistinguishable from angina pectoris is perhaps more common with partial absence of the pericardium. Moreover, the frightful potential of syncope and sudden death always exists in small defects of the pericardium and have been described. Transient incarceration of the left atrial appendage leading to mechanical restriction of cardiac activity may produce a serious fall in cardiac output and subsequent syncope. Two cases of sudden death have been reported, with strangulation of the heart as a result of herniation through a left pericardial foramen. A mechanical restriction of cardiac activity or strangulation would not be expected to occur if complete absence of the left pericardium exists. These possible ominous sequelae of partial defect argue for its surgical correction. Complete deficiency, on the other hand, would not require surgical intervention.

Cardiac enlargement may be more apparent than real, with complete absence of the left pericardium as a consequence of the leftward displacement of the heart. Clinically, this may be manifested as a forceful apical impulse beyond its normal boundary. The slow heart rate associated with complete heart block in this case may have influenced actual enlargement by dint of the increased stroke volume, with subsequent increased diastolic length of the myocardial fibers.

The life span of most patients with absent pericardium is probably not seriously compromised. The occurrence of pleuro-pericarditis as a complication of this congenital defect has been found in approximately 75 per cent of cases.

REFERENCES

CO$_2$-O$_2$ PNEUMOMEDIASTINOGRAPHY WITH POLY TOMOGRAPHY

The original technic of CO$_2$ pneumomediastinography has been modified by the injection of O$_2$ as contrast material after injecting CO$_2$ to dissect the subfacial planes of the mediastinum. Polytomograms of the mediastinum are then made for detailed demonstration of the anatomy.

CO$_2$-O$_2$ pneumomediastinography has been performed in this fashion in 64 instances of suspected bronchogenic carcinoma. This resulted in 87.5 percent technically successful examinations with no significant morbidity or mortality. It is emphasized that pathologic processes, grossly similar but histologically dissimilar, cannot be differentiated by this method.


GASTROESOPHAGEAL REFLUX AND HIATAL HERNIA

Gastroesophageal reflux secondary to cardiosophageal incompetence, with or without hiatal hernia, is a significant factor in the etiology of esophageal and respiratory tract complications. Cinefluorography of the esophagus has proved useful in diagnosis and evaluation of surgical therapy in patients with reflux. Indications for surgery include clinically asymptomatic patients resistant to medical treatment who demonstrate significant gastroesophageal reflux. Reconstruction of the gastroesophageal angle based on the technique of Belsey is a marked improvement over the modified Allison technique in restoring cardiosophageal competence.