Congenital Pericardial Defect Associated with Tetralogy of Fallot

Herniation of Normal Lung into the Pericardial Cavity

By Florencio A. Hipona, M.D., and Andrew B. Crummy, Jr., M.D.

Congenital pericardial defect is a rare anomaly, which was first described by M. Columbus in 1559.1, 2 Four hundred years later the first clinical diagnosis was reported by Ellis et al. In 1961, 108 cases were compiled from the literature and since then we have located reports of five more.3-6

Chang and Leigh1 reported that 67 per cent of the defects were left sided, and in three instances bilateral defects were present. Approximately three fourths of the defects were incomplete so that a foramen was present in the pericardium. The incidence of pericardial defects reported in male subjects is three times that in female.

The purpose of this communication is to record two cases of foramen-type pericardial defects. These are the first to be reported in association with tetralogy of Fallot. One case is unique in that normal lung had herniated into the pericardial cavity through the defect in the right side.

Case Reports

Case 1

A 6½-year-old white boy was seen in the Cardiac Clinic because of a heart murmur and slight circumoral cyanosis that had been initially noted at two weeks of age. Easy fatigability and dyspnea on exertion were first observed at 2½ years. These were gradually progressive to the point of dyspnea after climbing six to eight steps. The patient began to squat at 4½ years.

Physical examination revealed a ruddy-cheeked boy with bilateral inguinal hernias. The chest was symmetrical and the lungs were clear. The area of cardiac dullness was normal. There were a thrill and a loud systolic murmur along the upper left sternal border. The second sound in the pulmonic area was markedly diminished.

Electrocardiogram showed right axis deviation and right ventricular hypertrophy. Cardiac catheterization demonstrated a bidirectional shunt, predominantly right-to-left, at the ventricular level and marked right ventricular hypertension with an infundibular chamber.

Chest roentgenograms revealed a normal-sized heart with prominence of the lower anterior cardiac chamber indicative of right ventricular enlargement. The pulmonary artery segment was slightly concave. The pulmonary vascular pattern was normal. There was no evidence of left-sided enlargement and the aorta was of normal size with the arch to the left. An azygos lobe was present. A striking finding was the presence of a broad crescent of normal lung extending into the right superior mediastinum (fig. 1).

Selective right ventricular angiogram (fig. 2) demonstrated increased trabeculation of the right ventricle. There was a moderate-sized infundibular chamber bordered proximally by localized infundibular stenosis and distally by pulmonic valvular stenosis. Minimal early opacification of the aorta in the dextroangiogram indicated a right-to-left ventricular shunt. In addition, a left-to-right ventricular shunt was shown by a mixing defect in the right ventricular outflow tract in the dextroangiogram phase and reopacification of the pulmonary artery during the levoangiogram phase. The herniated portion of the right lung into the mediastinum was again identified.

At surgery a pericardial defect was found at the right posterolateral aspect superiorly. This permitted herniation of part of the normal right upper lobe into the pericardial sac. The right ventricle was enlarged with marked hypertrophy, especially in the infundibular area. Pulmonic valvular stenosis was present. The ventricular septal defect was closed with a Teflon patch after pulmonic valvulotomy and infundibular resection. The patient died two days later.

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132
CONGENITAL PERICARDIAL DEFECT

Figure 1
Case 1. Posteroanterior and right lateral films of the chest show a normal-sized heart with fullness anteriorly indicating right ventricular enlargement. The pulmonary vascular pattern is normal. An azygos lobe is present. Arrows outline the portion of the normal right upper lobe which has herniated into the pericardial cavity.

The findings at autopsy were infundibular stenosis, bicuspid stenotic pulmonary valve, ventricular septal defect, right ventricular hypertrophy (tetralogy of Fallot after open repair); interruption of continuity of the accessory conus branch of the right coronary artery; myocardial necrosis of the right ventricular wall near the apex; pericentral congestion and necrosis of liver; congenital defect in the right side of pericardial sac with herniation of lung parenchyma.

Comment. The herniation of lung into the pericardial cavity was evident in the roentgenograms. The superior vena cava and aorta distinctly bordered the herniated lung, which therefore assumed the shape of the space between these two structures.

Case 2
The patient was first seen at the age of 5½ years, December 1949. A murmur had been present since birth. Easy fatigability, squatting, and cyanosis were apparent following moderate exercise.

Physical examination showed an obese, short, white girl with dorsal scoliosis, internal strabismus, and cyanotic nail beds and lips. Her manner and speech suggested mental retardation. A grade-III/IV systolic murmur was heard best along the upper left sternal border.

Radiologic study in January 1950 showed the heart to be normal in size with a concavity in the region of the pulmonary artery segment. The pulmonary vasculature was diminished. There was fullness in the region of the right ventricle. The

Figure 2
Case 1. Right ventricular angiogram in the left posterior oblique position demonstrates stenosis in the region of the ostium infundibuli and the pulmonic valve with formation of an infundibular chamber. The right-to-left ventricular shunt is shown by early opacification of the aorta. The herniated portion of the right lung is outlined by arrows.
aortic arch was left sided. The radiographic appearance substantiated the clinical impression of tetralogy of Fallot.

A left Blalock procedure was performed in December 1951. At that time the left atrial appendage protruded through a circular defect in the left pericardium. The defect was closed with parietal pleura. The patient died 19 days later. Permission for necropsy was denied.

Discussion

Embryology

The consensus is that congenital pericardial defects are due to maldevelopment of the pleuropericardial membrane. Southworth and Stevenson 7 as well as Sunderland and Wright-Smith 8 have discussed the theories of embryogenesis in detail, but the exact mechanism remains unsettled.

Interference with the development of the membrane by premature atrophy of the left duct of Cuvier has been given as an explanation for the preponderance of left-sided defects. 1, 2 Others have postulated that herniation of an abnormal lung bud through the pleuropericardial foramen may result in the anomaly. 9 Several cases with a lung cyst in the region of the defect offer evidence in favor of this theory. 9, 10 Our first case would indicate that the lung need not be abnormal.

Congenital pericardial defects may occur alone or in association with other congenital anomalies. Table 1 1, 7, 8 shows that the combination of congenital pericardial defect and tetralogy of Fallot has not been reported.

Clinical Aspects

The incidental discovery of complete absence of the pericardium at operation or autopsy indicates that its presence is not essential for good cardiac function. 11, 12 Experimental evidence corroborates these clinical observations. 11

The cases in this report, as most others, had no symptoms attributable to the pericardial defect. Ellis et al., 2 however, reported two cases in which no other cause for chest pain was found. Hering et al. 4 described a case of chest pain relieved by enlargement of the defect in which the left atrial appendage became incarcerated. Three cases of acute compromise of cardiac function due to herniation of the heart resulting in sudden death have been described. 5, 8, 13

Radiologic Findings

The diagnosis may be made on plain chest roentgenograms. One of two conditions is necessary for it to be apparent: herniation of the heart or lung and pneumopericardium secondary to pneumothorax.

Ellis et al. 2 made the diagnosis when pneumopericardium occurred secondary to spontaneous pneumothorax. In their other case of an absent left pericardium, the diagnosis was suggested in plain films by shift of the heart to the left with exaggeration of the three distinct convexities of the left cardiac margin, namely, the aortic knob, the pulmonary artery segment, and the left ventricle.

Herniation through a foramen type defect may be seen on the chest roentgenogram. 3 The portion of the heart that herniates most commonly is the left atrial appendage. This may occasionally resemble a mediastinal mass and present a diagnostic problem. 4

Extension of lung beyond its normal confines into the mediastinum is considered mediastinal herniation. The usual type results in displacements of the heart or other mediastinal structures. This situation was thought to be present in our first case. In retrospect, the lung herniation did not occur in the usual manner; rather, it is into the pericardial cavity bordered by the aorta and superior vena cava. No reports of such herniation have been published.

Table 1

<table>
<thead>
<tr>
<th>Associated Cardiovascular and Pulmonary Anomalies</th>
<th>No. of Cases</th>
</tr>
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<tbody>
<tr>
<td>Patent ductus arteriosus</td>
<td>4</td>
</tr>
<tr>
<td>Bifid heart</td>
<td>2</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>2</td>
</tr>
<tr>
<td>Tricuspid insufficiency</td>
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</tr>
<tr>
<td>Bicuspid aortic valve</td>
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<tr>
<td>Bronchogenic cysts</td>
<td>6</td>
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<tr>
<td>Aberrant pulmonary lobe</td>
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</table>

* Our cases.
CONGENITAL PERICARDIAL DEFECT

Summary
Congenital pericardial defect is a rare anomaly with a total of 115 cases in the literature. The two cases discussed are the first reported to be associated with tetralogy of Fallot. The only known case of herniation of normal lung through a pericardial defect into the pericardial cavity is described.

References

Our Incomplete Knowledge of Cardiac Hypertrophy
There are many cases of hypertrophy, and of great hypertrophy of the heart, in which during life and after death no source of increased work can be discovered. The degree of enlargement found in many hearts in which there is a valve defect, such as aortic regurgitation or mitral stenosis, is sometimes out of all proportion to the apparent increase of burden. There is still much that remains to be explained; it is clear that there must be hidden sources of increased work, or the conclusion that increased work is the cause of hypertrophy needs revision.—Sir Thomas Lewis. Diseases of the Heart. New York, The Macmillan Company, 1933, p. 106.
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