Congenital Deficiency of the Pericardium*

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The parietal pericardium may be incompletely formed at birth, resulting in partial absence or deficiency of this membrane. As an isolated anomaly, the condition is rare. The subject was thoroughly reviewed by Ellis and associates in 1959. Less than 120 validated cases have been reported since that of Baillie in 1793. Despite this rarity, the condition is of interest to the clinician and radiologist because it may simulate more serious cardiac pathology. It may be associated with defects of thoracic structures or with other forms of congenital heart disease, and it may be a cause of unusual thoracic symptoms. Prior to 1936, congenital absence of the pericardium had been recognized only at necropsy. Since then, 29 cases have been diagnosed antemortem, including 21 at operation and eight by the demonstration of pneumopericardium after pneumothorax.

In this presentation, seven additional cases of congenital absence of the pericardium are reported. These cases demonstrate the association of the anomaly with other defects of thoracic structures, the ease of diagnosis by means of diagnostic pneumothorax, and the unique electrocardiographic findings in patients with large, left-sided defects of the parietal pericardium.

Material

The records of the Mayo Clinic include histories of six patients in whom thoracotomy revealed partial absence of the parietal pericardium and the history of one patient in whom such a deficiency was diagnosed by the demonstration of pneumopericardium after an artificial pneumothorax. There were six males whose ages ranged from three to 57 years and one female who was nine months old. The pericardial deficiency was associated with a ventricular septal defect in one patient and with patent ductus arteriosus in another. Two other patients had evidence of pulmonary disease, and three were studied because of suspected heart disease. One of these was found to have a large diaphragmatic hernia. In these five patients, no evidence of intrinsic cardiac disease was found.

Case Reports

Case 1

This patient was examined when he was 15 years old because of recurrent bouts of fever and hemoptysis during the preceding one and one-half years. Bronchographic studies revealed bronchiectasis in the left upper lobe. A left upper lobectomy was carried out. At the time of operation in 1945, a large defect was noted in the left side of the pericardium "through which a considerable portion of the heart could be visualized." After the operation, recovery was uneventful.

Case 2

A 57-year-old man underwent thoracotomy for carcinoma of the left lung. The left side of the pericardium was found to be absent. The right side was palpated and found to be intact. A left pneumonectomy was done, after which the heart could be rotated freely in the left side of the chest. Atrial fibrillation appeared as a transient dysrhythmia three days after operation. The patient was examined two years later and had a history consistent with angina pectoris which had developed one and one-half years after the pneumonectomy.

Case 3

This patient was first examined when he was 29 years old because of a six-year history of paroxysmal dysrhythmia. He had been in the United States Navy for 13 months without evidence of illness. Roentgenograms of the chest revealed that the heart was positioned mainly in the left hemithorax (Fig. 1A). The electrocardiogram (Fig. 4) showed right-axis deviation and clockwise rotation. A diagnosis of congenital defect of the pericardium was considered.

The patient was next examined when he was 36 years old. At this time, he complained of
burning epigastric distress, particularly during the night, fullness after meals, and frequent regurgitation of food. He continued to have paroxysmal dysrhythmia, one episode of which had been noted to be atrial fibrillation. Roentgenographic examination of the stomach revealed a large diaphragmatic hernia (Fig. 1B). Trans-thoracic repair of this hernia was undertaken. At operation, the pericardium was found to be almost completely lacking, except for a small vestige on the right. The hiatal hernia was unusual in that the stomach was not covered by a peritoneal sac, indicating that this was a part of a congenital peritoneal defect. The hernia was repaired and recovery was uneventful.

CASE 4

When this patient was 17 years old, he had been told that he had an enlarged heart that later returned to normal size. As an infant, he had two attacks of an influenza-like syndrome with fever and cough. Later that year, he moved to an area where coccidioidomycosis was endemic and he had "pneumonia." When he was 28 years old, he had subjective dyspnea and a wheezing sensation localized in the low retrosternal area when prone. This disappeared when he sat upright. A chest roentgenogram taken later that year was interpreted as showing cardiac enlargement, and he was referred for further study. After studies were carried out, exploratory thoracotomy was done and showed adhesive pleuritis over the left lung. The parietal pericardium was found to be absent on the left side, and the heart had been drawn by the adhesive pleuritis into the left chest. After decortication of the lung, the heart assumed an almost normal position.

CASE 5

A 22-year-old man had been told two years previously that his heart was enlarged. He had had no physical complaints. Cardiac auscultation was normal. Roentgenograms of the chest showed lateral displacement of the heart without sternal depression (Fig. 2). An electrocardiogram (Fig. 4) showed right-axis deviation and an rSr' complex in lead V-1 of normal duration. A provisional diagnosis of congenital deficiency of the pericardium was made. Because a definitive diagnosis was desired, an artificial pneumothorax was induced. Air (450 ml.) was introduced through a needle in the left fifth intercostal space in the mid-axillary line while the patient was lying on his right side. Air was demonstrated between the inferior surface of the heart and the diaphragm and in the retrosternal space and outlined the right side of the parietal pericardium (Fig. 1). The procedure was well tolerated.

CASE 6

This patient had a congenital heart lesion which was first noted when she was six weeks old. A murmur was heard, and blueness of the lips occurred with crying. She had frequent episodes of pneumonitis, grew slowly, and was underweight. A diagnosis of patent ductus arteriosus was made, and thoracotomy was carried out when she was nine months old. During the operation, an oval-shaped defect located on the left side and measuring 6 x 4 x 4 cm. was noted.

![Figure 1A, (Case 3): Posteroanterior view shows levo-displacement and elongation of heart, long and prominent pulmonary artery segment, and absence of apical fat pad. Figure 1B: Roentgenogram made during upper gastrointestinal examination shows moderate-sized esophageal hiatal hernia.](http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/21438/ on 04/19/2017)
CONGENITAL DEFICIENCY OF THE PERICARDIUM

Figure 2A, (Case 5): Posteranterior view shows shift of heart to left and unusual, elongated left heart border. Pulmonary artery segment is prominent and elongated. No apical fat pad is visible. Figure 2B: Lateral projection.

in the pericardium. The left auricular appendage was observed to herniate freely through the defect. The aortic arch was normal. The ductus was divided. An examination of the patient when she was six years old showed her to be apparently normal.

CASE 7

This patient had been considered to be normal at birth. A cardiac murmur had been noted when he was two months old, and subsequent investigation demonstrated ventricular septal defect and pulmonary stenosis. He had had a few episodes of bronchitis, but appeared to be well otherwise. At three years of age, he underwent surgical repair of the cardiac malformation. At operation, the patient was found to have a small congenital defect in the pericardium in the region of the left pulmonary artery. The ventricular septal defect was closed, the valvular pulmonary stenosis opened, and the infundibular pulmonary stenosis resected. The child was doing

Figure 3A, (Case 5): Anteroposterior view with patient supine made after 450 ml. of air was introduced into left pleural space. Air is seen separating heart from left diaphragm. Right side of pericardium and overlying pleura are seen in region of right cardiophrenic angle separating right lung and air in the pericardium. Figure 3B: Lateral view with patient supine utilizing horizontal beam after introduction of the air. In deficiency of the left side of the pericardium, the heart falls far away from the anterior chest wall and diaphragm. (Compare with upright lateral Fig. 2B).
well and had no cardiac symptoms when he was reexamined at age six years.

**DISCUSSION**

**Anatomic and Embryologic Features.**—The common defect of the parietal pericardium occurs on the left and is of variable size. When small, the defect lies superiorly, revealing the left pulmonary artery and left atrial appendage. When the defect is large, the free wall of the left ventricle may be uncovered or the entire cardiac apex may lie free in the left pleural space. In this series of seven patients, the defect was left-sided in all and large in five of the six who were operated on. These findings are in accord with those of Ellis and associates' who, in a review of the world literature, found 66 instances (65 per cent) of left-sided defects in 99 patients. Of these, 48 (73 per cent) had large defects. In the same study, Ellis and collaborators' found that males predominated (73 per cent of 81 patients for whom sex was reported). Six of the seven patients in the present study were males. The higher incidence of left-sided defects and of males with congenital pericardial deficiency is unexplained.

The common defect of the parietal pericardium occurs as the result of failure of development of the right or left pleuropericardial membrane. These paired structures which close the primitive pleuropericardial canals by the seventh week of embryonic life arise as folds on the lateral margins of the transverse septum and body wall. They are associated with the cardinal veins which pass ventromedially in the transverse septum to join the sinus venosus. During normal embryologic growth, the pleuropericardial folds, with the cardinal veins, move toward the midline and fuse with pre-esophageal mesenchyme, thus closing the pleuropericardial canals and completing the formation of the parietal pericardium. At the same time, the cardinal veins undergo change. The right cardinal vein persists as the superior vena cava while the left atrophies. Perna" postulated that premature atrophy of the left cardinal vein would result in a left-sided pericardial defect. This theory of origin of pericardial defects finds support in the high incidence of left-sided, as compared to right-sided, defects.

**FIGURE 4A:** Electrocardiogram in cases 1 through 5 from above downward.
CONGENITAL DEFICIENCY OF THE PERICARDIUM

On embryologic grounds, an association of pericardial and diaphragmatic defects may be anticipated. In a few cases, deficiency of the parietal pericardium may be the result of the failure of a portion of the transverse septum to develop and may be associated with a complex anomaly, including defects of diaphragm, chest wall, and abdominal viscera. A simpler defect was observed by Ladd in 1936 in the first instance of pericardial deficiency to be recognized during life. Ladd's patient, a two-year-old girl, had a hernia of the left lateral portion of the diaphragm, probably because of the failure of the pleuropitoneal membrane to develop. At repair of this defect through an abdominal incision, the pericardial deficiency was observed. One patient in the present series (case 3) had an esophageal hiatal hernia. Surgical repair of this was done with a transthoracic approach that afforded an excellent view of the heart and which demonstrated the deficiency of the pericardium. At the same time, it was noted that there was no peritoneal sac covering the herniated portion of the stomach. This observation supports the impression that a developmental defect might be responsible for the complex of pericardial and diaphragmatic abnormalities observed in certain patients.

Electrocardiographic Features — The electrocardiographic findings in congenital absence of the pericardium may be deserving of more attention. In this group of five patients with large, left-sided defects, the electrocardiographic findings were remarkably similar and, although not grossly abnormal, they were unusual (Fig. 4). Common to the group was right-axis deviation and displacement of the transition zone to the left in the precordial leads. This combination of changes is unexpected in the more common forms of congenital and acquired heart disease. In association with the radiologic abnormalities, the electrocardiogram led to a working diagnosis of congenital pericardial defect in two patients (cases 3 and 5).

A review of the literature reveals little concerning the electrocardiographic findings in congenital absence of the pericardium. A single tracing has been published and it is of limited value, for it is from a two-year-old child with a patent ductus arteriosus and reflects the hemodynamic state related to the patent ductus. Other authors, in reporting isolated cases, have noted “incomplete right bundle branch block,” “clockwise rotation and borderline left axis deviation,” “right axis deviation,” or “normal curves.”

**Figure 4B:** Electrocardiogram in cases 2 through 5 from above downward.
Although regarded as nonspecific, the observed electrocardiographic changes likely reflect the abnormal position of the heart within the thorax. Since heart position is determined in part by the size of the pericardial defect, small defects should have no distinguishing electrocardiographic features, whereas large defects may be associated with electrocardiographic findings resembling those reported herein.

**Radiologic Features**—Ellis and his associates, in 1959, reported the first two cases of congenital pericardial deficiency in which the diagnosis had been suggested on the basis of the conventional chest roentgenogram. Both of these cases were considered examples of complete absence of the left side of the parietal pericardium. The roentgenographic features in these cases were: (1) a shift of the heart to the left, and (2) an unusual cardiac silhouette with a somewhat elongated left heart border and a long, prominent, and sharply demarcated pulmonary artery segment.

In six of the seven cases comprising our series, large left-sided defects were present; in five of these, the roentgenographic features described by Ellis and his associates were evident. Also noted in six of the seven patients was absence of the normal apical epicardial fat pad. The presence of a band of radiolucency between the base of the heart and the diaphragm was noted in three patients, was not present in three, and was obscured by pleural adhesions in one patient. These findings probably are a logical consequence of complete absence of the left side of the pericardium. Although these features can in no way be considered diagnostic of congenital pericardial deficiency, the diagnosis is to be strongly considered when they are observed.

Partial or foraminal type defects in the left side of the pericardium would not be expected to result in abnormality of the cardiac silhouette unless a portion of the heart was herniated through the defect. Kavanagh-Gray and associates, in 1961, reported a case in which the left atrial appendage was herniated through the left pleuropericardial foramen. The chest roentgenogram in this case showed an unusual, protruding shadow along the left heart border, which on angiocardiography proved to represent the herniated left atrial appendage. To our knowledge, no abnormal roentgenographic findings have been recorded in other more rare types of congenital pericardial deficiency, such as deficiency of the right side of the pericardium or isolated defects in the diaphragmatic portion of the pericardium.

In patients with absence of a large portion of the left side of the pericardium, the sternopericardial ligament may be lacking, so that with the supine position the heart is permitted to fall away from the anterior chest wall and diaphragm. This abnormal mobility of the heart may be readily demonstrated by means of a roentgenogram made with a horizontally directed beam and the patient in the supine position. A plain x-ray film in supine lateral projection might provide a simple screening procedure in cases in which a large defect is suspected.

The demonstration of pneumopericardium after the induction of the pneumothorax may be regarded as diagnostic of congenital pericardial deficiency. For this examination, 400 to 500 ml. of air are introduced into the pleural space. Many positional changes may be necessary to permit filling of the pericardium prior to roentgenography. With a horizontal beam, filming is carried out in the anteroposterior and lateral projections with the patient supine and with the patient in the lateral decubitus position. For examination in the left lateral decubitus position, one film should be made with the patient in the straight decubitus position and one with the patient in a slightly right posterior oblique position in relation to the film. This latter position may prove most informative since the right heart border is not superimposed on the spinal column. Anteroposterior supine tomograms may also be useful in demonstrating air separating the base of the
heart from the dome of the diaphragm. Upright postero-anterior and lateral films after the introduction of air are evidently of little value.

A pneumothorax may not always yield diagnostic information since, in very small pericardial defects, air may not gain entrance to the pericardial cavity. Furthermore, in rare instances, pericardial adhesions may be present and may prevent accumulation of air within the pericardium.

Angiocardiography has little positive value in the diagnosis of congenital pericardial deficiency, but may be indicated in some cases in order to rule out other cardiac abnormalities.

**Surgical Considerations.** — Pericardial deficiencies often are incidental findings at thoracotomy and ordinarily require no specific measures for correction. This is particularly true of major deficiencies or complete absence of the pericardium. Only when there is partial absence of the pericardium which may lead to herniation" or strangulation of a portion of the heart, usually the left atrium or its appendage, should consideration be given to specific surgical treatment. The adjacent pleura has been used successfully as a patch to cover the defect after excising the appendage."

Another surgical approach to prevent herniation and possible strangulation of the heart is the conversion of a small defect to a large one by excision of some of the existing pericardium. Either approach would seem to be logical and effective.

**Clinical Features.**—Congenital defect of the pericardium ordinarily is not associated with symptoms. Palpitation and tachycardia have been reported, but these are not specific symptoms, being found with many kinds of heart disease or with no disease at all. There are case reports of sudden death in which the atrial appendage or the apex of the heart prolapsed through the defect and strangulated,"17,18,31

**Summary**

Deficiency of the parietal pericardium was demonstrated in seven patients at the Mayo Clinic. The defect was left-sided in all seven and large in five of the six patients who underwent thoracotomy. Diagnosis was established in one case by the demonstration of pneumopericardium after artificial pneumothorax was made for that purpose. Six of the group were male. In five patients without heart disease, the electrocardiogram revealed right-axis deviation and late development of the R deflection in precordial leads. The electrocardiogram may be of value in the diagnosis of congenital pericardial deficiency.

**Resumen**

En 7 pacientes observados en la clínica Mayo se comprobó solución de continuidad en el pericardio parietal. Este defecto asentaba en lado izquierdo en todos y era de tamaño considerable en 5 de los 6 casos en que se practicó la toracotomía.

El diagnóstico se estableció en un caso mediante la inducción de neumotorax, que dio lugar a un pneumopericardio.

Seis de los pacientes eran del sexo masculino. En cinco de ellos, no afectos de cardiopatías, el ECG reveló dextro-rotación del eje eléctrico y mas tarde el desarrollo de la onda R en la derivación precordial. El electrocardiograma puede ser de valor en el diagnóstico de los defectos congénitos del pericardio.

**Zusammenfassung**

Ein Fehlen des parietalen Peri cardiablattes konnte am Material der Mayo-Klinik in 7 Fällen nachgewiesen werden. Bei allen 7 Fällen handelte es sich um einen linkssitzigen Defekt, und er war erheblich groß bei 5 der 6 Patienten, die mit Thorakotomie behandelt wurden. Die Diagnose wurde ermittelt in einem Fall durch den Nachweis eines Pneumopericards, nachdem ein Künstlicher Pneumothorax aus diesem Zweck
PULMONARY ALVEOLAR PROTEINOSIS — REPORT OF A CASE WITH SPONTANEOUS RESOLUTION

This diffuse pulmonary infiltrate was first described by Rose, Castieman and Liebow in 1958. In the case reported by the authors, the condition was accidentally found on a routine chest x-ray and the diagnosis was confirmed by pulmonary biopsy. The lesion resolved spontaneously within one year. A detailed study of the case is presented and the literature is reviewed.


REPORT OF 26 CASES OF SUB-STERNAL GOITER

The authors report 26 cases of substernal goiter which were removed surgically. In 15 cases, the goiters were truly substernal, in ten a small part of the gland was within the thorax and in one case there was a second thyroid gland deeper within the thorax. Radiosotope studies were not very helpful in localizing the lesions. In almost all of the cases, the cervical approach was satisfactorily used. Three patients died, one with carcinoma of the thyroid, another with a benign 750 gram goiter and the third, the only endo-thoracic goiter.