Congenital pericardial defects

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Four cases of congenital defect of the pericardium are described; three had complete absence of the left pericardium and one had a partial defect. One of the patients developed acute pulmonary oedema in the immediate post-operative period following repair of a sinus venous defect. This complication was thought to be directly related to the pericardial defect. The functions of the pericardium are discussed. The clinical features of the different types of pericardial defect are described. The diagnosis is usually made from the radiographic appearances. Electrical alternans has been noticed in one patient. This is probably due to the increased mobility of the heart. These defects are not completely benign, as has been suggested before. They can cause disabling symptoms, as in case 3, and may be directly responsible for death.

Congenital defects of the pericardium were first reported by Columbo (1559) and Baillie (1793). Until recently the condition was described as an incidental post-mortem finding and aroused the interest mainly of anatomists and embryologists. Ellis, Leeds, and Himmelstein (1959) reported the first case of complete absence of the left pericardium diagnosed radiologically. The clinical importance of pericardial defects has not been sufficiently stressed. Many authors regard the condition as benign (Southworth and Stevenson, 1938; Moore and Shumacker, 1953; Tabakin, Hanson, Tampas, and Caldwell, 1965; Nasser, Feigenbaum, and Helmen, 1966) in spite of the fact that these defects may produce symptoms and in some cases may be directly responsible for death. In this paper four patients with congenital pericardial defects are described. The main clinical, radiographic, and electrocardiographic features are pointed out, and the possible haemodynamic effects are discussed.

CASE REPORTS

CASE 1 A man aged 37 was admitted to the Brompton Hospital on 7 July 1966 after having had a routine chest film taken. He stated that he had been advised to avoid vigorous exercise following a chest radiograph taken at the age of 14. He could play games including tennis until two years before admission, but admitted that he was slightly more short of breath than other people of his age. On examination he was fit, and the pulse was regular with a rate of 75 per minute. The jugular venous pressure was normal. The apex beat was shifted to the left and downwards, while the trachea was central. He had a pulmonary ejection murmur, grade 2 in intensity, with a split second sound. The pulmonary component of the second sound moved normally on respiration.

Chest radiographs (Figs 1 and 2) showed displacement of the cardiac shadow to the left with a central trachea. The main pulmonary artery was prominent, and the space between it and the aorta was wide. The lateral film showed displacement of the posterior border of the heart backwards. An electrocardiogram showed sinus rhythm with a PR interval of 0.18 second. There was right axis deviation with a mean frontal QRS vector of 110° and the pattern of right bundle-branch block.

Cardiac catheterization was performed on 11 July. This showed normal pressures with a left-to-right shunt at atrial level. The pulmonary to systemic flow ratio was 2.5:1. Dye dilution curves suggested preferential shunt from the right pulmonary veins. Five days after cardiac catheterization, while being driven home by his wife, he suddenly felt himself urinate and then noticed that he could not move his right arm or speak. This lasted for 10 minutes but recurred a few hours later for a period of half an hour, with complete recovery. There was no evidence of any residual neurological damage. These episodes were thought to have been due to paradoxical cerebral embolism from the vein used for cardiac catheterization. He was treated with anticoagulants.

Repair of the atrial septal defect was performed on 2 August through a right anterolateral thoracotomy with transverse division of the sternum, as the heart was markedly shifted to the left. The internal mammary vessels were tied and divided. The pericardium on the right side was normal. On opening the pericardium it was realized that the left side of the pericardium was completely absent with a com-
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**FIG. 1.** Case 1. Chest radiograph showing displacement of the cardiac shadow to the left and prominence of the pulmonary artery shadow.

**FIG. 2.** Case 1. Lateral chest film showing displacement of the posterior border of the heart backwards.

Mon pleuropericardial cavity on the left side. The right phrenic nerve was normal in position, but the left phrenic nerve ran behind the sternum and was mistaken for the left internal mammary vessels and divided before opening the pericardium. It ran from the anterior chest wall on to the upper surface of the central tendon of the diaphragm, then turned sharply to the left to supply the left dome of the diaphragm. The anterior surface of the heart was formed by the right atrium, the ventricles lying in the para-vertebral gutter. Any attempt at restoring the heart to its normal position resulted in an immediate fall in blood pressure. There was a sinus venosus defect with anomalous upper and middle right pulmonary veins. A small secundum atrial septal defect was also present. These defects were repaired under cardio-pulmonary bypass. During the immediate post-operative period the patient was well. He was fully conscious with signs of a very good cardiac output. The arterial oxygen saturation was 99% and the venous oxygen was 75%. The venous pressure was 2 cm. of water above the sternal angle. During the next six hours he was transfused with 900 ml. of blood; his central venous pressure remained low (4 cm. of water). However, he suddenly developed severe pulmonary oedema. At that stage he was still being artificially ventilated through an endotracheal tube. Large
amounts of pink froth were aspirated from the tube. The lungs were stiff with a high inflation pressure. The arterial oxygen saturation dropped to 60% in spite of positive pressure respiration using 100% oxygen. Five hundred millilitres of blood were withdrawn from the circulation and positive pressure respiration was continued. Pulmonary oedema steadily improved. Positive pressure respiration had to be continued for a long time, because five days after operation he developed bilateral severe lung infection with *Pseudomonas pyocyanea* which was resistant to treatment. He died on 9 September 1966. Necropsy showed a large tracheo-oesophageal fistula at the site of the balloon of the tracheostomy tube. The repair of the septal defects and the anomalous pulmonary veins was satisfactory, with no evident narrowing of any pulmonary vein. All cardiac valves were normal.

Case 2 A boy aged 6 was admitted to the Brompton Hospital on 21 January 1952. He complained of a persistent cough and expectoration of a large amount of yellow sputum. He was a well-nourished child with grade 2 clubbing of the fingers. The trachea was central, but the apex beat was markedly shifted to the left. The movements of the left lower chest were poor, and there were coarse crepitations over the left lower lobe. A chest radiograph (Fig. 3) showed a marked shift of the cardiac shadow to the left. The main pulmonary artery was prominent, and the space between it and the aorta was widened. The lower third of the left border of the cardiac shadow was ill defined.

A left lower lobectomy was performed on 7 March. The basal segments of the lower lobe were atelectatic. The lingula appeared normal. The left pericardium was completely absent with a common left pleuropicardial cavity. Over the apex of the heart there was a patch of opaque epicardium. The left phrenic nerve ran extrapericardially, lateral to the internal mammary vessels.

Post-operatively he made an uneventful recovery. He was seen recently, when he stated that he had no chest symptoms apart from vague discomfort in the left chest and occasional 'cramp' in the left lower chest on laughing. Examination showed a marked shift of the heart to the left and a pulmonary ejection murmur. An electrocardiogram showed sinus rhythm with a rate of 70. The PR interval measured 0.2 second. The mean frontal QRS vector was +110° with a pattern of partial right bundle-branch block.

Case 3 A boy aged 17 years was admitted to the North Middlesex Hospital on 6 August 1967. He complained of severe sharp pain in the left side of the chest, which had started three years previously while he was playing football. The pain was precipitated by exercise, lying on the left side, and sometimes by lying flat. He was moderately incapacitated by the pain, which had been getting progressively worse. He also complained of palpitations which accompanied the pain. He denied any fainting sensation. On examination he was normally developed and the pulse was regular and equal on both sides. The jugular venous pressure was normal. The left side of the chest was flattened and he had an accessory nipple below and medial to the normally situated left nipple. The apex beat was displaced to the mid-axillary line. He had a moderately loud pulmonary ejection murmur. The pulmonary component of the second sound moved normally. The chest radiograph (Fig. 4) showed all the features of an absent left pericardium. The electrocardiogram showed sinus rhythm with a mean frontal QRS vector of approximately +60°. Some of the leads showed total electrical alternans which involved the frontal QRS vector and possibly the P vector, and was most pronounced in aVL and aVF (Fig. 5). The vector shift occurred every third beat and involved two beats before returning to the original position. On exercise the electrical alternans occurred every second beat, and this was not accompanied by any change in heart rate. The electrical alternans was not associated with pulsus alternans. The duration of QRS was 0.1 second and showed a pattern of partial right bundle-branch block with a small R′ in V1. Cardiac catheterization was performed on 25 September and showed normal right-sided pressures with no evidence of any shunts. The diagnosis of an absent left pericardium was confirmed by inducing a left pneumothorax (Fig. 6, a and b). Thoracoscopy was performed and showed complete absence of the left pericardium with no visible pericardial edges. Exploratory thoracotomy was performed on 24 October; this showed no pleural or pericardial adhesions. The left pleuropicardium was absent apart from a small crescentic fold at the upper angle. There was no pericardial shelf and no chance of the heart getting incarcerated, as it was completely free to move in the common pleuropicardial cavity. The chest was closed in layers. The patient made a satisfactory recovery. He was seen three months later, when he still complained of the same pain which prevented him from finding a suitable occupation.

Case 4 A man aged 22 years was admitted to hospital on 13 October 1966. Four weeks before admission he had coughed up blood-stained sputum for four days. A routine chest film had been reported on as showing an abnormal shadow one year previously. Examination of the chest and cardiovascular systems showed no abnormal signs. A chest radiograph (Fig. 7) showed an opacity in the region of the left atrial appendage. An electrocardiogram was within normal limits. Cardiac catheterization showed a pulmonary artery pressure of 23/5 mm. Hg and a mean pulmonary wedge pressure of 2 mm. Hg. Pulmonary angiograms were done, but failed to opacify the region of the abnormal shadow. Bronchoscopy showed no endobronchial abnormality. Thoracotomy was performed on 20 October. There were a few adhesions between the lung and the
FIGS 3 and 4. Cases 2 and 3 (respectively). Chest radiographs showing all the features of an absent left pericardium.
mediastinal pleura around a pleuro-pericardial defect in front of the upper part of the hilum of the lung. The defect was circular and measured about 1.5 in. (38 mm.) in diameter. The left atrial appendage was moderately enlarged and protruded through the defect. The adhesions were divided and the chest was closed.

Post-operatively the patient made a smooth recovery.

DISCUSSION

Congenital pericardial defects are rare; only one case was encountered in a series of 14,000 necropsies at the Johns Hopkins Hospital (Southworth and Stevenson, 1938). These anomalies can be classified into:

1. Those due to deficiency in the development of the pleuropericardium
   
   (a) Partial or foramen type
   
   (b) Complete absence of the pericardium (pleuropericardium) on one side (usually the left) or very rarely both sides.

2. Those due to deficiency in the development of the septum transversum. This is usually associated with a defect in the central tendon of the diaphragm and results in a rare type of diaphragmatic hernia, in which the abdominal contents herniate into the pericardial cavity (Gross, 1946; Keith, 1910; Kadin, 1939).

Pleuropericardial defects, whether partial or complete, are commoner on the left side. This is probably explained by the fact that the developing pleuropericardial membranes are intimately related to the ducts of Cuvier. The left duct of Cuvier normally atrophies while the right one persists as the superior vena cava. Premature atrophy of the left duct of Cuvier may predispose to the development of a pleuropericardial defect. Southworth and Stevenson (1938) reviewed all the previously reported cases of congenital pericardial defects and found that complete absence of the left pericardium accounted for 76% of these cases. However, recently partial defects are being reported more frequently (Rusby and Sellors, 1945; Fry, 1953; Jones, 1955; Dimond, Kittle, and Voth, 1960; Hering, Wilson, and Ball, 1960; Chang and Leigh, 1961; Kavanagh-Gray, Musgrove, and Stanwood, 1962; Bruning, 1962; Duffle, Moss, and Maloney, 1962; Tucker, Miller, and Jacoby, 1963; Williams, 1963; Hipona and Crummy, 1964; Mukerjee, 1964; Baker, Schlang, and Ballenger, 1965). This is probably explained by

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FIG. 5. Case 3. Electrocardiogram showing total electrical alternans which is most pronounced in lead I, aVL, and aVF and occurs every third beat. On exercise the alternans occurred every second beat.
FIG. 6 (a and b). Case 3. Chest radiographs after induction of left pneumothorax, showing air in the pericardium and outlining the right parietal pericardium as a linear shadow.
the fact that most of the earlier cases were recognized at necropsy, and it would be easier to miss a partial defect, especially when small, during routine post-mortem examination. Pericardial defects affect men more commonly than women, the ratio being 3:1.

Complete absence of the left pericardium presents a different clinical picture from partial defects. Many patients complain of pain in the left hypochondrium; this is usually precipitated by exercise and lying on the left side, and can be incapacitating, as in case 3. The cause of this pain is not known; but the most probable cause is kinking of the great vessels. Sometimes the pain is associated with a feeling of faintness (Fisher and Ehrenhaft, 1964). Sinus bradycardia is sometimes present. The heart is shifted to the left, but not the other mediastinal structures. A pulmonary ejection murmur and accentuation of the pulmonary component of the second sound are almost always present. The electrocardiogram usually shows marked right axis deviation and the pattern of right bundle-branch block. These signs suggest the diagnosis of atrial septal defect, which is not uncommonly present in association with complete absence of the left pericardium (as in case 1 in this paper and the patients reported by Fisher and Ehrenhaft (1964) and Tabakin et al. (1965)). This should always be excluded by cardiac catheterization. Total electrical alternans, as observed in case 3, has not been described before in association with absent left pericardium. The alternation of both the QRS and P vectors is probably due to the increased mobility of the heart. This is supported by the fact that the only other condition known to produce total electrical alternans is pericardial effusion (Traut, 1950; Colvin, 1958; Littmann and Spodick, 1958; Lawrence and Cronin, 1963), where there is increased freedom of movement of the heart. The diagnosis of absent left pericardium is usually made by the characteristic radiographic appearances. These include a marked shift of the cardiac shadow to the left with some enlargement of heart size. The pulmonary artery
shadow is prominent and the space between it and the aorta is widened with interposition of lung between the main pulmonary artery and the aorta. The left border of the heart shows two well-marked convexities formed by the pulmonary artery and ventricular mass. The left atrial appendage is not seen on the P.A. film due to rotation of the heart towards the left. The upper border of the pulmonary artery shadow forms an acute angle with the mediastinal shadow. The lower part of the left border of the heart is ill-defined. This seems to be a constant sign which has not been described before and is probably due to increased mobility of the apex. The lower border of the heart is elongated and flattened, with interposition of lung between it and the diaphragm. The lateral view shows displacement of the posterior border of the heart backwards; this is produced by the apex of the heart, which lies in the paravertebral gutter. Tomograms confirm interposition of lung between the main pulmonary artery and the aorta. In doubtful cases the diagnosis can be proved by inducing a left pneumothorax and taking films in the upright, Trendelenburg, and left lateral positions.

Absent left pericardium is not uncommonly associated with other congenital anomalies. These include secundum atrial septal defect (Fisher and Ehrenhaft, 1964), sinus venosus defect with partial anomalous pulmonary venous drainage (case 1 in this paper; Tabakin et al., 1965), extralobar pulmonary sequestration (Hamilton, 1961), and diaphragmatic hernia (Ladd, 1936).

In patients who have an absent left pericardium the heart lies free in the common pleuroperticardial cavity and the pericardium ceases to be a confined space. Several functions have been given to the pericardium. These include:

1. Prevention of overdistension of the heart (Kuno, 1915; Berglund, Sarnoff, and Isaacs, 1955; Holt, Rhode, and Kines, 1960);
2. Protection of the lungs against pulmonary oedema in cases of left ventricular failure by interfering with right ventricular filling (Berglund et al., 1955);
3. Reducing the tendency to regurgitation through the atrio-ventricular valves in cases of raised atrial pressures (Berglund et al., 1955);
4. Prevention of spread of infection from the lung and pleura to the serous pericardium and heart;
5. Maintaining the heart in the position of optimum function.

Nelemans (1940) showed that the pericardium is formed of interwoven elastic and collagenous fibres. The former allows the pericardium to stretch up to a point, after which collagenous fibres resist any further expansion. Isaacs, Berglund, and Sarnoff (1954) studied the pressure volume curve of the pericardium, and showed that increase in volume produced little change in pressure up to a point, after which any small increase in volume resulted in a steep rise of pressure. Holt et al. (1960) showed that a rise of venous pressure produced by acute hypervolaemia was accompanied by a rise of intrapericardial pressure; thus the actual filling pressure to which the myocardium was subjected during diastole was less than the venous pressure. This showed that the pericardium prevented overdistension of the ventricles during diastole in cases of raised venous pressure. Carleton (1929) showed that removal of the pericardium in cats produced permanent enlargement of the cardiac shadow, which he thought was due to dilatation and not hypertrophy. In one of the patients reported by Tucker et al. (1963), wide splitting of the left pericardium resulted in 'definite enlargement of the heart shadow'. Kuno (1915) found that removal of the pericardium in the heart-lung preparation tended to produce cardiac dilatation and lower filling pressure. He concluded that a heart in the absence of the pericardium was already in danger when subjected during diastole to a venous pressure of one-third or one-half that found under normal conditions. All these experiments showed that the heart can function normally in the absence of pericardium so long as it is not subjected to high venous pressures.

The protective function of the pericardium against pulmonary oedema is suggested by the experiments of Berglund et al. (1955). They showed that in dogs with a high left atrial pressure, produced by partial clamping of the aorta and increasing the blood volume, opening the pericardium produced a fall of right atrial pressure and a rise of left atrial pressure to dangerous levels. This suggested that in the presence of the pericardium the dilated failing left ventricle interfered with filling of the right ventricle and so protected the lungs against pulmonary oedema. This was further supported by their study of ventricular function curves on partially clamping the aorta and so increasing the load on the left ventricle. In the presence of the pericardium this produced depression of both left and right ventricular function curves. After excising the pericardium partial clamping of the aorta resulted in depression of the
left ventricular function curve, but the right ventricular function curve remained unchanged. The same mechanism may explain the occurrence of pulmonary oedema in case I in this paper.

Southworth and Stevenson (1938) found that in 27% of previously described cases of congenital defects of the pericardium, death was associated with pleuropericarditis. With the use of antibiotics this ceased to be a problem, although pleuropericardial adhesions were not uncommon in cases of defects of the pericardium, which suggested that the fibrous pericardium prevented spread of infection to the contents of the pericardial cavity.

Partial defects of the pleuropericardium are usually small (foramen type) and situated behind the phrenic nerve in the uppermost part of the pericardium in front of the hilum of the lung. They are more common on the left side. The left atrial appendage enlarges and herniates through the defect. This produces the characteristic radiographic appearance. In the absence of mitral valve disease, enlargement of the left atrial appendage suggests the presence of a partial defect of the pericardium. Chest pain has been described in some cases (Hering et al., 1960; Duffie et al., 1962). This may be due to partial strangulation of the herniated left atrial appendage. The enlarged atrial appendage increases the risk of systemic emboli (Williams, 1963) and should therefore be excised. The association of a foramen type defect and a bronchogenic cyst has been described (Rusby and Sellors, 1945; Jones, 1955; Mukerjee, 1964). Rusby and Sellors suggested that the defect could be the result of herniation of part of the abnormal lung bud medial to the duct of Cuvier, and so interfere with the development of the pleuropericardial membrane. Herniation of a normal lung into the pericardium has been described (Hipona and Crummy, 1964). Intrapericardial bronchogenic cysts with intact pericardium have also been reported (Leagus, Gregorksi, Crittenden, Johnson, and Lepley, 1966).1

Moderate-sized pericardial defects are dangerous, as the ventricles may herniate through the defect and become strangulated, resulting in sudden death, as in the cases reported by Boxall (1887), Sunderland and Wright Smith (1944), and Bruning (1962). These defects are fortunately extremely rare. The mechanism of strangulation is identical to cases described after intrapericardial pneumonectomy with excision of a patch of pericardium (Yacoub, Williams, and Ahmad, 1968).

We should like to thank Mr. R. Bates, Mr. Raymond Hurt, and Dr. B. Wells for their permission to publish details of patients who were under their care.

REFERENCES


1In a personal communication, Barrett has told us of a young man, who was symptomless, and who, on screening, was found to have a large pulsating opacity in the left upper mediastinum. At thoracotomy this proved to be a cyst, containing clear fluid and lined by columnar epithelium. It arose by a pedicle from the aortic arch and was removed. The pericardium above the ventilices was deficient and the heart could easily be lifted out of the bag in which it lay. The patient recovered uneventfully and, as far as is known, had had no further trouble.
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