**CASE REPORT**

**Congenital absence of pericardium: two cases and a comprehensive review of the literature**

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**ABSTRACT**

Congenital absence of pericardium (CAP) is a rare condition, generally asymptomatic or paucisymptomatic, nevertheless sporadic cases complicated by sudden death are described. CAP can be diagnosed by CT and MRI. It is classified as total or partial, and partial defects are divided into left defects and right defects. Interestingly, several articles highlight the correlation between CAP and some anatomical lung abnormalities, such as presence of lung parenchyma between the main pulmonary artery and ascending aorta, lung parenchyma between the base of the heart and left hemidiaphragm, and lung parenchyma between the proximal ascending aorta and right pulmonary artery.

**CASE PRESENTATION**

The first case is a 66-year-old male, presented to the Emergency Department of Azienda Ospedaliera di Cremona complaining of atypical chest pain and palpitations. His medical past history was unremarkable. ECG revealed right axis deviation, a right bundle branch block, and no ST elevation. The troponin I value was mildly elevated, 40.1 ng l$^{-1}$ (normal range: 0–34.2 ng l$^{-1}$). We suspected an acute coronary syndrome, the patient was underwent to cardiac catheterization, which revealed no obstructive coronary artery disease of the left main coronary, but the right coronary artery was not detected. In order to exclude an aortic dissection or an anomalous origin of the right coronary, a CT angiography was carried out. CT showed a marked displacement of the heart in the left hemithorax. The pericardium was only partially visualized (Figure 1), lung tissue was evident between the ascending aorta and the main pulmonary artery, and between the base of the heart and the left hemidiaphragm (Figure 2a and b). The findings were considered signs of congenital absence of the pericardium.

The second patient was a 49-year-old female, brought to the Emergency Department because of a road accident. Her chest X-ray was interesting, showing leftward position of the heart, and hyperlucency between the aorta and main...
The pericardium is partially covering the right cardiac chambers and is absent over the left ventricle (indicated by arrows). Pulmonary artery due to interposition of lung (Figure 3). The review of a previous chest CT showed a left pericardium defect.

**DISCUSSION**

Congenital absence of the pericardium is an uncommon cardiac defect estimated to occur in 1 in 10,000–14,000 people, and is usually discovered incidentally, e.g. during operations, autopsies and radiological studies.1–4 Most patients are asymptomatic, but atypical chest pain, dyspnoea and palpitations are common symptoms.1–3,5 Patients with CAP have a life expectancy comparable to that of the general population, but life-threatening complications, such as incarceration of left atrial appendage, ventricular herniation and torsion of the great vessels are described in the literature.3,6

Up to 30% of pericardial defects are associated with other cardiac abnormalities, including bicuspid aortic valve, mitral valve stenosis or prolapse, tetralogy of Fallot, atrial septal defect, or persistent ductus arteriosus.2,5,6 In addition, a variety of extra-cardiac malformations can also be associated with pericardial defects. These abnormalities include bronchogenic cysts, pulmonary sequestration, VACTER syndrome, diaphragmatic hernia, or hepatic hemangioendothelioma.2,5,6

Six categories of pericardial defects have been reported: total absence, right-sided defects (complete or partial), left-sided defects (complete or partial), and diaphragmatic defects.5

Left-sided defects are the most commonly reported, involving up to 70–80% of cases of pericardial defects, whereas right-sided pericardial absence is much rarer than left-sided defects and was reported in 4–17% of cases. Finally, total absence of the pericardium is the rarest of all defects, seen in only 9% of cases reported in the literature.3,6

In case of absent left pericardium, CT typically shows some indirect signs, such as abnormal presence of lung parenchyma in the usually lung-free aortopulmonary space and interposition of lung tissue between the left hemidiaphragm and the base of the heart.1–3,7–9

A direct sign of left CAP is represented by blocked view of the left pericardium.

In case of absent right pericardium, CT may show, as indirect sign, an abnormal presence of lung parenchyma in the space, usually lung-free, between the ascending aorta and the right pulmonary artery, and an anomalous cardiac contour due to right appendage, atrium or ventricle herniation.3,5,10–12 A direct sign of right CAP is represented by a blocked view of the right pericardium.

In case of a total absence of the pericardium, a combination of the previously described indirect signs may appear.1 A marked displacement of the heart into the left hemithorax may be present in total and left CAP.

**Figure 1.** The pericardium is partially covering the right cardiac chambers and is absent over the left ventricle (indicated by arrows).

**Figure 2.** (a). Interposition of lung tissue between the aorta and main pulmonary artery (L). (b). Interposition of lung tissue between the diaphragm and the base of the heart (L).
Total defects are of little clinical importance, but partial defects, although rarely, may be the cause of sudden death, therefore the detection of this malformation is clinically important, and the recognition of the pulmonary anatomical abnormalities related to CAP may be useful clues to suggest and support the diagnosis.

We compared the most common described lung abnormalities associated with CAP (Table 1) with data from our experience (Table 2). We detected 1 case of left absence of pericardium in 12,288 individuals examined with chest CT.

Moreover, we identified two cases with lung interposition between the ascending aorta and main pulmonary artery, one case characterized by CAP, specifically a left defect (Figures 1, 2a and b). The other case involved a patient with no pericardial defect and with a history of mediastinal emphysema caused by complications that arose during the insertion of a right internal jugular central venous catheter (Figure 4). At the time of insertion of the central venous catheter, the presence of the aortic-pulmonary pericardial recess was evident (Figure 4a and b), but no longer clearly evident at the CT check about 2 years later (Figure 4c and d). We speculated on the possible cause of the pericardial recess changes, considering the hypothesis that the post-procedural mediastinal emphysema could have caused a regional fat necrosis, and we also hypothesize that major thoracic traumas might cause similar anomalies.11

In our experience, no case with interposition of pulmonary parenchyma between the ascending aorta and right pulmonary artery was detected.

Finally, the presence of pulmonary parenchyma between the inferior side of the heart and the left hemidiaphragm was found in several patients, one with left absence of pericardium (Figures 1, 2a and b), and all the others characterized by dorsal kyphosis (Figure 3), cause of displacement of the heart into the left hemithorax and interposition of pulmonary parenchyma between the base of the heart and left hemidiaphragm. In conclusion, we suggest taking into consideration the lung anomalies associated with CAP, although not exclusive of this condition, to support its diagnosis, and to properly recognize a total or partial defect, because of the potential complications in partial defects.

Table 2. Indirect signs of CAP: frequency from our data

| Indirect signs of CAP: frequency from our data |
|-----------------------------------------------|
| Lung parenchyma between the main pulmonary artery and ascending aorta: |
| 2 in 1,288 cases, |
| • 1 case of partial left CAP a |
| • 1 case of post-procedural event, without CAP |
| Lung parenchyma between the base of the heart and left hemidiaphragm: |
| 10 in 1,288 cases, |
| • 1 case of partial left CAP a |
| • 9 cases with dorsal kyphosis, causing heart displacement with lung interposition between heart and left hemidiaphragm, without CAP |
| Lung parenchyma between the proximal ascending aorta and right pulmonary artery: |
| 0 in 1,288 cases |

CAP, congenital absence of pericardium.

*aSame patient showing left CAP, with the evidence of both lung between the main pulmonary artery and ascending aorta, and lung between the base of the heart and left hemidiaphragm.
Learning Points

1. Congenital absence of pericardium is a rare condition, usually discovered incidentally. This entity can be diagnosed by CT and MRI.
2. It is widely known the correlation between congenital absence of pericardium and some anatomical lung abnormalities, such as:
   - lung parenchyma between the main pulmonary artery and ascending aorta,
   - lung parenchyma between the base of the heart and left hemidiaphragm,
   - lung parenchyma between the proximal ascending aorta and right pulmonary artery.
3. It is useful taking into consideration these lung anomalies to support the diagnosis of congenital absence of pericardium, and to properly recognize a total or partial defect, because of the potential complications in partial defects.

Conflicts of Interest:
no relationship with industry and financial associations within the past 2 years that poses a conflict of interest in connection with the submitted article exists.
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