Lost in space – left lateral cardiac displacement due to an unusual cause

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We present a case of an incidental finding of congenital absent pericardium (CAP). A 69–year-old patient was admitted to the intensive care unit because of septic shock of unknown cause. The patient's history was remarkable for alcoholic liver disease with cirrhosis and lung emphysema. The chest X-ray (CXR) revealed an enlarged and displaced cardiac silhouette of atypical shape without signs of congestive heart failure. Electrocardiogram (ECG) showed right axis deviation, leftward displaced transition zone in the precordial leads and significant changes of QRS axis and R wave amplitude between admission and follow-up. Echocardiographic images were hard to obtain and atypical lateral views were required. Computed tomography of the chest revealed cardiac displacement into the left thoracic cavity. Congenital absent pericardium (CAP) was assumed. Shortly after, the patient died of lung failure. Autopsy confirmed pneumonia as cause of death and incidentally revealed a complete isolated absence of the pericardium and a displaced heart to the left.

CAP was first described by Realdus Colombus (1559) [1]. The aetiology of this anomaly is an incomplete development of the transversal septum and the pleuropericardial folding in the early embryonic stage [2]. The prevalence is unknown; various authors have described isolated forms in surgical or pathological series of 0.0001% to 0.044%. One third of all cases were associated with other cardiac lesions, like atrial septal defect, bicuspid aortic valve and tetralogy of Fallot. Patients can present asymptomatic or with atypical chest pain, largely non-exertional and with postural dependency. Partial defects can lead to cardiac herniation with shock or sudden death. Clinical examination may show lateral displacement of the heart. ECG often displays a partial right bundle block pattern and a deviated transition zone to the left. CXR can show a levoshift with loss of the right heart border. Often lung tissue is interposed between main pulmonary artery and aorta. Although echocardiographic examination requires unusual views and is difficult to interpret because of cardiac hypermobility leading to abnormal swinging and ventricular septal motion, the change of the position of the heart from left side to supine position is a key finding for this diagnosis by echocardiography. CXR combined with magnetic resonance imaging are key to establish the diagnosis. In patients with debilitating symptoms pericardial reconstruction is a curative option [3].

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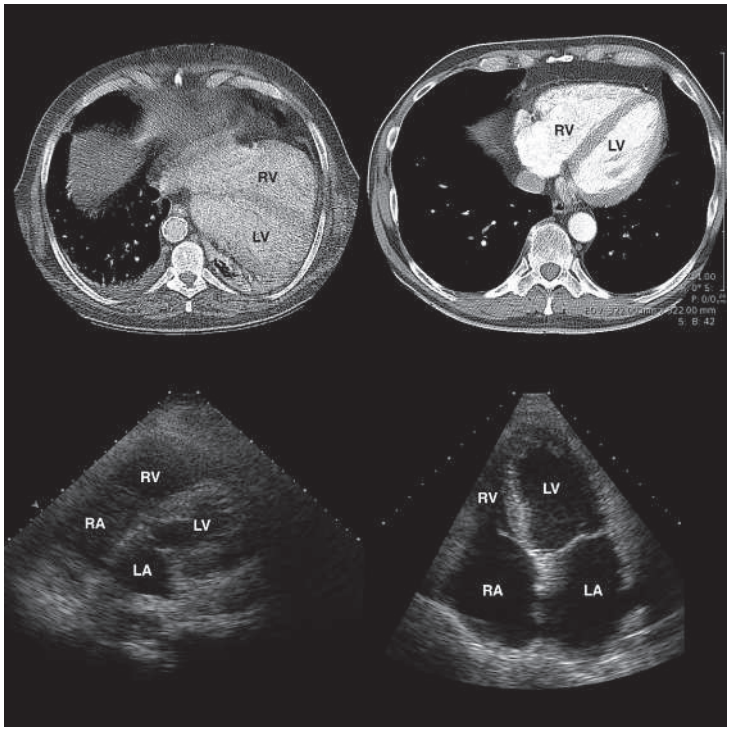
Figure 1
Cardiac displacement to the left (above left) compared with normal cardiac position (above right). Unusual echocardiographic findings (below left) with the transducer held in usual left lateral position for apical four-chamber view compared with a normal echocardiographic orientation (below right) [4]. LA left atrium, LV left ventricle, RA right atrium, RV right ventricle.



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