

Congenital absence of the pericardium

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Published online: 13 April 2011

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A 71-year-old male presented with atypical chest pain and dilatation of the right ventricle on echocardiography and was referred for cardiac magnetic resonance imaging (CMR) for suspected arrhythmogenic right ventricular cardiomyopathy. CMR showed marked posterior and leftward displacement of the heart, with the apex extending to the left lateral chest wall. Spin echo imaging showed thin pericardium around the atria and great vessels, but no

pericardium was detected around the ventricles (Fig. 1). Lung tissue was interpositioned between the heart and the left hemidiaphragm (Fig. 2). Both ventricles had normal morphology and function.

Patients with congenitally absent pericardium may be asymptomatic or may present with chest pain or dyspnoea [1]. Electrocardiography may show right bundle branch block or clockwise rotation. Chest X-ray may show the left lateral position of the heart and the abnormal interposition of lung tissue between the aorta and pulmonary artery or between the inferior myocardial wall and the left hemidiaphragm. On echocardiography, the right ventricle may appear enlarged, as the beam traverses the ventricle closer to the basis of the heart and tangentially [2].

The diagnosis can be confirmed by computed tomography or CMR by showing (partial) absence of the pericardium.

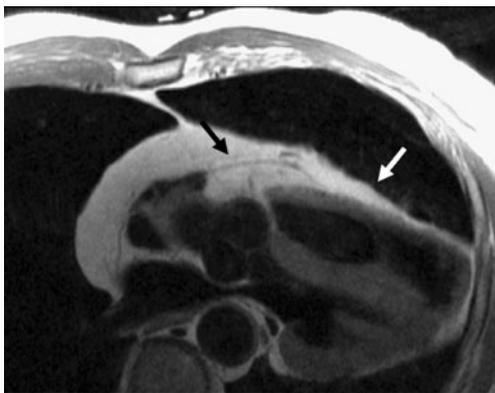


Fig. 1 Spin echo study showing pericardium around the right atrium (*black arrow*), but lack of pericardium around the right ventricle (*white arrow*)

Electronic supplementary material The online version of this article (doi:10.1007/s12471-011-0111-7) contains supplementary material, which is available to authorized users.

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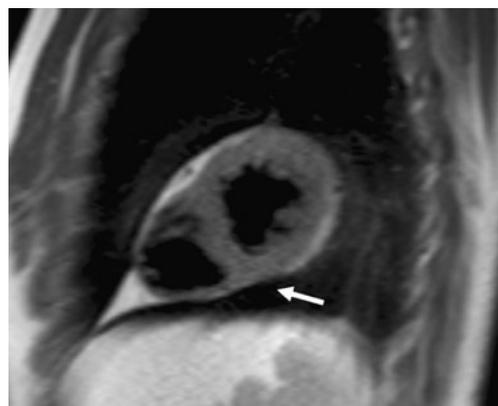


Fig. 2 Still frames from steady state free precession cine loop showing lung tissue interposition between the inferior myocardial wall and the diaphragm (*white arrow*). Original cine loops published online (Movie 1)

CMR may show excessive cardiac mobility. In incomplete defects, a rim of residual pericardium may cause a left ventricular crease, with (fatal) herniation as a rare potential risk, for which preventive closure has been suggested. Complete forms only need intervention in case of symptoms [3, 4].

Consent

Written informed consent was obtained from the patient.

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