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Echocardiography

Congenital Complete Absence of the Pericardium: A Multimodality Imaging Diagnostic Approach

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Because of an occasional finding of extreme leftward and posterior rotation of the cardiac apex on chest radiograph, a 16-year-old man was referred to our hospital for further investigation with magnetic resonance imaging (1.5 Tesla MRI scanner; General Electric Medical System, Waukesha, Wisconsin, USA) to look for pericardial tissue and to rule out other causes of cardiac displacement. His medical history, family history, and physical examination were unremarkable. Electrocardiography demonstrated right axis deviation and an incomplete right bundle branch. Magnetic resonance imaging confirmed the heart displaced into the left hemithorax and showed the distinct pathognomonic feature of interposition of the lung in the usually lung-free space between heart and diaphragm, both in coronal and sagittal axis (Fig. 1). No pericardium was present over most of the heart. On the basis of these results, we suspected a complete congenital absence of the pericardium. Transthoracic echocardiography showed normal left and right ventricular function and no other structural heart disease; the parasternal long-axis view evidenced unusual acoustic windows with left ventricular posterior wall displaced toward a deeper thoracic location in end-diastole (movie clip 1). Short-axis view showed pendulum-like movement of the heart (movie clip 2). In the apical four-chamber view, cardiac hypermobility with swinging motion of the heart and teardrop appearance with bulbous ventricle and elongated atria were noted (movie clip 3). M mode examination demonstrated an apparent paradoxical septal motion with a flat systolic motion of interventricular septum (Fig. 2) assumed to be due to exaggerated anterior translational motion.

In this case, echocardiography permitted the distinction between partial and complete pericardial absence because these echocardiographic features are typical of the whole left and right pericardium absence[1,2] This distinction is

Conflict of Interest: None declared.

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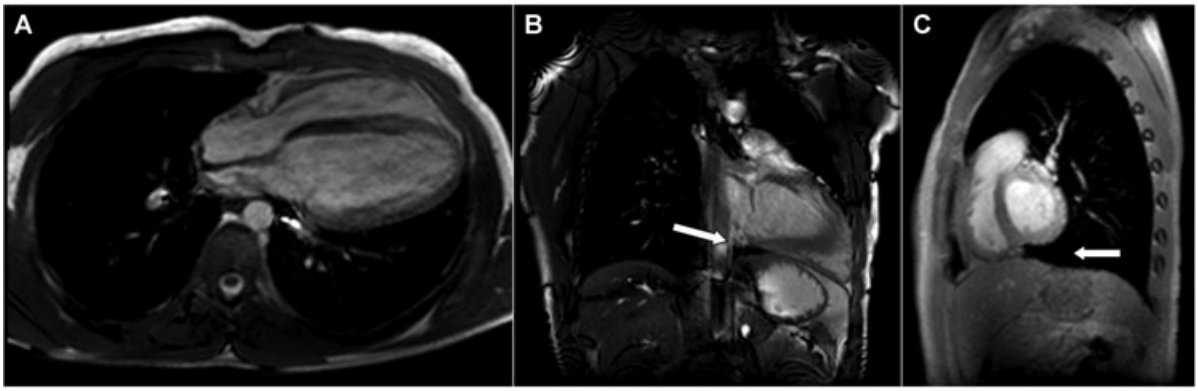

Figure 1. Magnetic resonance imaging: **A.** heart displaced into the left hemithorax and interposition of the lung between the heart and diaphragm (arrow) both in **B.** coronal and **C.** sagittal axis.

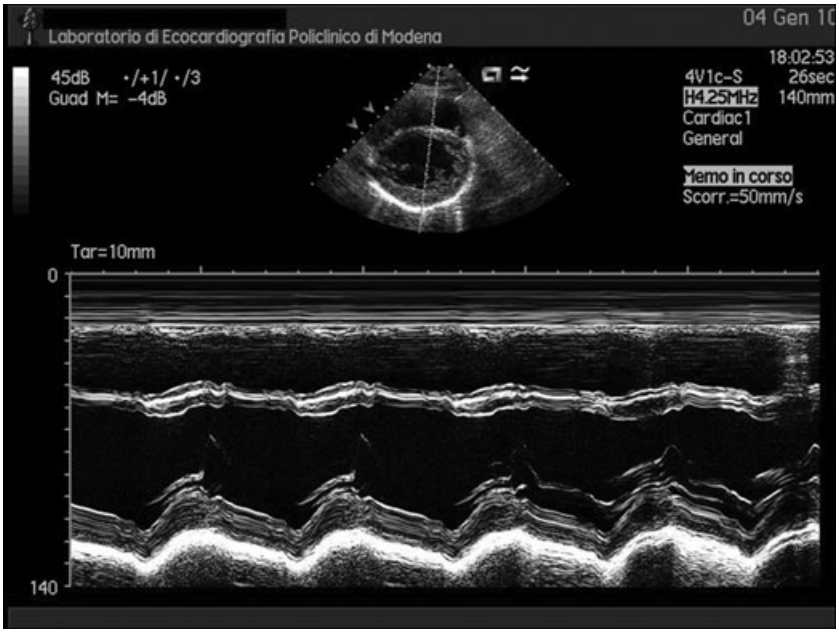

Figure 2. Flat systolic motion of interventricular septum.

clinically relevant since patients with partial congenital absence of pericardium are more at risk of herniation and morbidity. Importantly, a partial pericardial defect closure reduces the risk of complications and of severe outcomes. In contrast, complete absence of the entire pericardium usually has a benign prognosis and it requires no intervention,[3] so the patient was reassured and continued his activities without limitation.

References

1. Centola M, Longo M, De Marco F, et al: Does echocardiography play a role in the clinical diagnosis of congenital absence of pericardium? A case presentation and a systematic review. *J Cardiovasc Med* 2009;10:687–692.
2. Connolly HM, Click RL, Schattenberg TT, et al: Congenital absence of the pericardium: Echocardiography as a diagnostic tool. *J Am Soc Echocardiogr* 1995;8:87–92.
3. Gatzoulis MA, Munk MD, Merchant N, et al: Isolated congenital absence of the pericardium: Clinical presentation, diagnosis, and management. *Ann Thorac Surg* 2000;69:1209–1215.

Supporting Information

Additional Supporting Information may be found in the online version of this article:

Movie clip 1. Parasternal long-axis view: unusual acoustic windows with left ventricular posterior wall displaced toward a deeper thoracic location in end-diastole.

Movie clip 2. Short axis view showed pendulum-like movement of the heart.

Movie clip 3. Apical four-chamber view: cardiac hypermobility with swinging motion of the heart, teardrop appearance with bulbous ventricle and elongated atria.

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