

CLINICAL VIGNETTE

Complete absence of the pericardium in a competitive athlete

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We report a case of a 16-year-old asymptomatic girl who was referred for a paediatric cardiology consultation due to a heart murmur detected during a regular check-up. The patient had no medically relevant background and played indoor football for a federated team three times a week. Physical examination was unremarkable except for a discrete systolic heart murmur. A 12-lead electrocardiogram showed poor R-wave progression at the precordial leads and right axis deviation (Fig. 1A). On chest X-ray, there was a levorotation of the heart, with no projection of the cardiac silhouette on the right side of the spine, an elongated left heart border, and radiolucency between the diaphragm and the heart (Fig. 1B). Transthoracic echocardiogram (TTE) in standard apical four-chamber view showed right ventricular dilation (Fig. 1C), and atypical lateroposterior views were necessary for better visualisation. TTE revealed exaggerated mobility of the heart, mild mitral regurgitation with no valvular prolapse, elongated ventricles, and normal biventricular systolic function. To clarify these findings, magnetic resonance imaging was performed which depicted teardrop-shaped heart (Fig. 2A), left posterolateral rotation of the heart with apex orientated towards the mid-axillary line, and interposition of lung parenchyma between the aorta and the main pulmonary artery (Fig. 2C) as well as between the inferior aspect of the left ventricle and the diaphragm (Fig. 2B). There was also protrusion of the left atrial wall on both sides of the descending aorta (Fig. 2D). No pericardial recesses were seen between the aortic root and the main pulmonary artery. The pericardium could not be observed in any of the sequences obtained. No other cardiac or vascular abnormalities were found. These results were consistent with the diagnosis of complete congenital absence of the pericardium. Given the usually benign course and complete absence of symptoms, we opted for a conservative approach, with periodic ambulatory observation. A 24-hour Holter electrocardiogram monitoring showed sinus rhythm without dysrhythmias, bradycardia runs, or right bundle branch block. We also performed an exercise stress test, which revealed no abnormalities. However, given the lack of evidence in these cases, we advised against maintaining high-intensity and contact sport activity at a competitive level. Congenital absence of the pericardium is a very rare cardiac anomaly. Patients are usually asymptomatic, although they can present with positional chest pain, syncope, or even sudden death in cases of partial defects due to cardiac strangulation. Our patient was asymptomatic, and her cardiac condition did not seem to limit her exercise tolerance. However, there is little evidence regarding the safety for these patients to perform competitive high-intensity and contact exercises. In the future, exercise cardiac magnetic resonance imaging might prove to be helpful in giving advice in rare conditions like this.

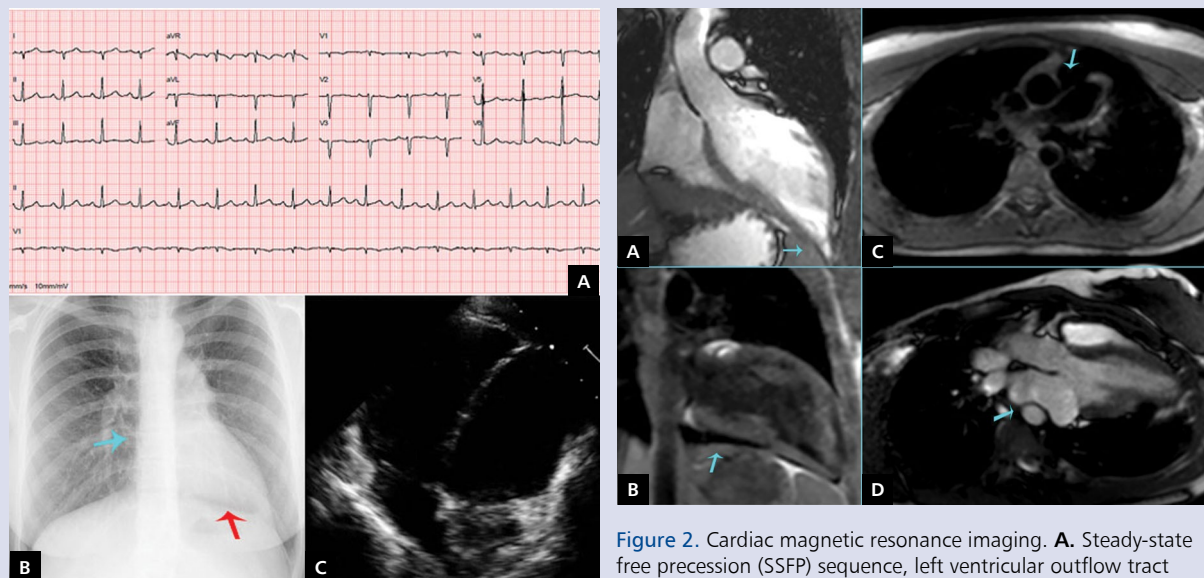


Figure 1. **A.** 12-lead electrocardiogram; **B.** Chest X-ray, postero-anterior view. Levorotation of the heart, with no projection of the cardiac silhouette on the right side of the spine (blue arrow), an elongated left heart border and radiolucency between the diaphragm and the heart (red arrow); **C.** Transthoracic echocardiogram, standard apical four-chamber view

Figure 2. Cardiac magnetic resonance imaging. **A.** Steady-state free precession (SSFP) sequence, left ventricular outflow tract view. Teardrop-shaped heart (blue arrow); **B.** Black blood sequence, two-chamber view. Radiolucency between the diaphragm and the heart (blue arrow); **C.** Black blood sequence, axial view of the great vessels. Interposition of lung parenchyma between the aorta and the main pulmonary artery (blue arrow); **D.** SSFP sequence, three-chamber view. Protrusion of the left atrial wall on both sides of the descending aorta (blue arrow)

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Conflict of interest: none declared

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