INTRODUCTION

Congenital pericardial defect is a rare and usually asymptomatic condition which is classified incomplete or partial. Up to 70% of cases consist of complete absence of left pericardium. The diagnosis may be challenging due to its low frequency and absence of correlation with any specific finding on the clinical examination. Cardiac magnetic resonance imaging is the gold standard imaging technique for the diagnosis confirming the absence of pericardium, although other indirect signs may be seen. In partial defects, surgery is the treatment option. We present an incidental finding of total agenesis of the left pericardium in an asymptomatic 16-year-old male diagnosed in a preoperative assessment of a bone fracture.

CASE REPORT

An asymptomatic and healthy 16-year-old male was referred to the cardiology clinic due to a heart murmur noted in a preoperative assessment of left ulnar fracture. Physical examination revealed the presence of pectus carinatum, an apical palpation deviated to left, and I/VI systolic murmur with normal heart sounds. In addition, there was a swinging heart noted in the right precordium. Twelve lead electrocardiogram (ECG) showed sinus rhythm (90 bpm) with right axis deviation, signs of right chambers enlargement, and the absence of R-wave growth in precordial leads [Figure 1]. Chest X-ray is described in Figure 2. Echocardiography (Philips iE33, 5-3 Hz) demonstrated a structurally and functionally normal heart with left heart deviation and no pericardial effusion. In addition, a standardized cardiac magnetic resonance imaging (MRI) (Philips Achieva 1.5T) was performed to complete investigations [Figure 3]. Given the anatomical distortion of the patient, the location of the interactive planes was very challenging. Both biventricular size and function and flow imaging were unremarkable. However, an extreme leftward shift of the heart was identified, with the apex pointing posteriorly and the right ventricle in close contact to the left anterior chest wall. In addition, axial fast imaging employing steady-state acquisition demonstrated...
According to what is described in the literature, all these clinical and imaging findings suggest the diagnosis of total agenesis of the left pericardium.

**DISCUSSION**

Congenital pericardial defect is a rare condition secondary to a disturbance of pleuropericardial membranes fusion on one or both sides during fetal life. It can be classified in partial or complete (being lungs and heart are in the same cavity) [1]. Up to 70% of cases consist on complete absence of left pericardium being rare in right side (17%), complete bilateral defect is less common (9%–13%).[1,2] The majority of patients are asymptomatic, and diagnosis may be challenging due to its low frequency and absence of correlation with any specific finding on the clinical examination. There is a higher prevalence in males (3:1), and no familial association has been identified. Associated congenital cardiopulmonary anomalies are found in 30%–50% of patients. In ECG right axial deviation, incomplete right bundle branch block pattern and leftward displacement of the transitional zone in precordial leads are seen.

On echocardiography, only indirect signs can be analyzed. Cardiac MRI is the imaging technique of choice. Normal pericardium is shown as a hypointense linear band between epicardial adipose tissue (hyperintense), myocardial tissue (medium intensity), and the surrounding adipose tissue around the heart. However, in 10% of the patients, there is a small amount of adipose tissue that hinders the visualization of the interposition of lung tissue between the aorta and pulmonary artery. Short-axis steady-state free precession (SSFP) sequence showed the portion of the posterior wall of the left ventricle (between arrows) which was presumed to be the pericardial defect. The remaining pericardium was seen as a low-intensity linear band (arrow heads).

Cine SSFP sequences depicted left lung interposition between left hemidiaphragm and the inferior side of the heart [Videos 1 and 2].
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Pericardium. Thoracoscopy is the technique of choice for confirmation.\(^1\)

Pericardium is easier to identify during the systole above the right ventricle. Thickness of normal pericardium ranges from 1.7 ± 0.5 mm to 1.2 ± 0.5 mm in diastole.\(^1\)

Not all pericardial recesses are seen on MRI, although some of them may be detected such as transverse pericardial sinus, dorsal to the ascending aorta, and oblique pericardial sinus, behind the left atrium.\(^1\) Other signs that can be shown are an abnormal heart position, bulging of the main pulmonary artery, and interposition of lung tissue between left hemidiaphragm and the inferior side of the heart and between great vessels, being the last two specific findings in complete left defect.\(^1\) A herniation of the heart through pericardial defect can also be seen.\(^1\)

Surgery is done in partial defects (to prevent the mechanical complications of the heart), when there are complications or in symptomatic patients.\(^1,2\) Surgical options could be: Patch closure of the pericardial foramen, enlargement of the defect, pericardiectomy, or pericardioplasty.\(^2\) The prognosis is good in the complete absence of pericardium, so no treatment is needed unless mechanical complications appear.

Our patient remains asymptomatic, and no other complementary study or cardiological intervention has been carried out.

**Declaration of patient consent**

The authors certify that they had all appropriate patient consent forms. In the form, the patient has given his consent for his images, and other clinical information to be reported in the journal. The patient understands that his name and initial will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

**REFERENCES**

1. Cuccuini M, Lisi F, Consoli A, Mancini S, Bellino V, Galanti G, et al. Congenital defects of pericardium: Case reports and review of literature. Ital J Anat Embryol 2013;118:136-50.

2. Shah AB, Kronzon I. Congenital defects of the pericardium: A review. Eur Heart J Cardiovasc Imaging 2015;16:821-7.