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Quantitative criteria for the diagnosis of the congenital absence of pericardium by cardiac magnetic resonance

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ABSTRACT

Background: Congenital absence of the left ventricular pericardium (LCAP) is a rare and poorly known cardiac malformation. Cardiac Magnetic Resonance (CMR) is generally used for the diagnosis of LCAP because of its high soft tissue contrast, multiplanarity and cine capability, but the diagnosis is usually made by only qualitative criteria. The aim of the present study was to establish quantitative criteria for the accurate diagnosis of LCAP on CMR.

Methods: We enrolled nine consecutive patients affected by LCAP (mean age 26 ± 8 years, 7 males), 13 healthy controls, 13 patients with dilated cardiomyopathy (DCM), 12 patients with hypertrophic cardiomyopathy (HCM) and 13 patients with right ventricular overload (RVO). All patients underwent CMR. The whole-heart volume was measured in end-systole and end-diastole. Whole-heart volume change (WHVC), was the systo-diastolic change of volume, expressed percentage of the end-diastolic volume. The angle of clockwise-rotation of the heart was also measured in the end-diastolic phase of the axial cine stack.

Results: The WHVC was significantly higher in LCAP (21.9 ± 5.4), compared to healthy subjects (8.6 ± 2.4, $p < 0.001$), DCM (7.1 ± 1.8, $p < 0.001$), HCM (9.3 ± 2.4, $p < 0.001$) and RVO (8 ± 2.4, $p < 0.001$). The clockwise-rotation was significantly higher in LCAP (76 ± 14°) than healthy controls (40 ± 11°, $p < 0.001$), DCM (41 ± 5°, $p < 0.001$), HCM (30 ± 6°, $p < 0.001$) and RVO (49 ± 8°, $p < 0.001$). WHVC had the highest sensitivity (100%) and specificity (100%) for diagnosing LCAP, using a threshold of >13%.

Conclusions: In LCAP the systo-diastolic WHVC was significantly higher than controls, DCM, HCM and RVO patients and resulted an optimal quantitative criteria for the diagnosis of LCAP.

1. Background

Congenital absence of the left pericardium (LCAP) is an uncommon cardiac defect that occurs as a consequence of premature atrophy of the left common cardiac vein (Cuvier vein), with insufficient blood supply to the pleuropericardium leading to its agenesis [1,2]. It is estimated to occur in 1:10,000–14,000 people and is three times more common in males [3]. This condition is sometimes identified with the alternative term "pericardial agenesia" but in the current manuscript we preferred the term "pericardial absence".

The LCAP is frequently isolated but it may also be part of more complex cardiac and extracardiac malformations [4] such as in VATER (vertebrae, anus, trachea, esophagus, and renal) syndrome, Marfan syndrome, and Pallister-Killian syndrome [5].

LCAP is more commonly found than right-side absence of pericardium [6–8] and more frequently associated with complications, because of the absence of anatomical barriers. Although the prognosis is usually benign, LCAP can lead to serious complications such as incarceration of cardiac tissue, myocardial ischemia, aortic

Abbreviations: LCAP, left ventricular congenital absence of the pericardium; CMR, cardiac magnetic resonance; HCM, hypertrophic cardiomyopathy; DCM, dilated cardiomyopathy; RCO, right ventricular overload; SSFP, steady-state free precession; FSE, fast spin echo; WHVC, systo-diastolic whole heart volume change; RV, right ventricle; LV, left ventricle; DTD, end-diastolic transverse diameter; STD, end-systolic transverse diameter; DLD, end-diastolic longitudinal diameter; SLD, end-systolic longitudinal diameter.

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dissection, recurrent pulmonary infections and valvular insufficiencies [7,9]

Even if echocardiography and chest X-ray studies may help to detect agenesis of the pericardium, the images may not be considered diagnostic because they do not directly show the pericardium and pericardial defects, but provide only indirect clues [9]. Cardiac magnetic resonance (CMR) is the preferred technique for the diagnosis of LCAP. CMR delineates the anatomical relationships between the heart, lung and mediastinum with high soft tissue contrast and multiplanar capability [9,10–12]. The demonstration of the presence of faded heart–lung interface because of the partial volume effect in fast-spin echo (FSE) images, together with the morphological and functional features showed in cine-images, are qualitative CMR findings in LCAP. However, the diagnosis of a pericardial defect is not always straight forward even on CMR, since in normal conditions the pericardium over the lateral side of the left ventricle, corresponding to the most frequent location of pericardial defects, is usually not well depicted because of a paucity of surrounding fat [13]. Moreover, the morphological and functional characteristics of LCAP are evaluated only qualitatively.

In the current study we evaluated different quantitative criteria for the diagnosis of LCAP and compared the effectiveness of these criteria for the differential diagnosis among LCAP, healthy controls and patients with other cardiac conditions, namely hypertrophic cardiomyopathy (HCM), dilated cardiomyopathy (DCM) and right ventricular overload (RVO).

2. Methods

2.1. Study population

From November 2003 to December 2014 we enrolled nine consecutive patients with a diagnosis of LCAP (7 males, mean age 26 ± 8 years) who underwent CMR. The diagnosis of LCAP was performed by the summation of clinical, electrocardiographic, chest X-ray and qualitative CMR data in all the subjects. Two patients underwent cardiac surgery for LCAP, directly confirming the diagnosis.

Thirteen healthy subjects (10 males, age 25 ± 8 years) were enrolled as control group. Also, 13 patients with a known diagnosis of DCM (12 males aged 31 ± 6 years), 12 patients with HCM (10 males, age 29 ± 6 years) and 13 patients with RVO (9 males aged 25 ± 8 years), were enrolled in order to test the quantitative criteria of LCAP diagnosis in comparison with models of, respectively, dilated and hypertrophic hearts. The diagnosis of HCM and DCM was based on the presence of standard CMR and clinical criteria [14,15].

The group of patients with RVO included five patients with anomalous pulmonary veins return, three patients with atrial septal defect with left-to-right shunt, two patients with severe pulmonary valve stenosis, two patients with severe tricuspid regurgitation, and one patient with post-embolic pulmonary hypertension.

2.2. Cardiovascular magnetic resonance (CMR)

CMR examination was performed using a 1.5-T Signa CVi scanner (GE, Milwaukee, Wisconsin) with a cardiac phased-array 8-channel coil. For the assessment of regional wall motion and left and right ventricular volumes and mass, cine images were used with a steady-state free precession (SSFP) pulse sequence in short-axis views (from atrioventricular valve plane to the apex, 8-mm slice thickness, no gap) and in axial views (from diaphragm to the entire outflow tract, 5-mm slice thickness, no gap). The following acquisition parameters were applied: 30 phases, 10 to 20 views per segment depending on heart rate, NEX 1, FOV 40 cm, a matrix of 224–224, a 45° flip angle, TR/TE equal to 3.5/1.5, and a bandwidth of 125 kHz.

For the evaluation of the interface between epicardial fat-pericardial layer and sub-epicardial myocardium, FSE echo images were acquired in the same short-axis views (8 mm slice thickness, no gap) and para-axial views (5 mm slice thickness, no gap) with the following parameters: NEX 1, FOV 40 cm, matrix of 256–256, a 90° flip angle, average TR/TE of 1791/41.5, and a bandwidth of 62.5 kHz; a double inversion-recovery preparation pulse was applied to suppress the boodpool signal. The same stacks of images were also reacquired using a fat saturation pulse to selectively null signals from fat.

2.3. Cardiac magnetic resonance data analysis

The standard qualitative CMR features on black blood FSE and in SSFP sequences were: (a) absence of the hypointense line of the pericardium surrounding the left ventricular walls, which separates the epicardial adipose tissue (of high signal intensity) or myocardial tissue (of medium signal intensity) from the high intense pericardial adipose tissue surrounding the heart; (b) a shaded heart–lung parenchyma transition due to the partial volume effect; (c) an abnormal extension of the main pulmonary artery and the left atrial appendage beyond the margins of the mediastinum; (d) interposition of lung tissue between the aorta and pulmonary artery or between the diaphragm and the base of the heart, (e) cardiac indentation at the location of the defect [5,16]. The functional CMR features were: (a) the leftward displacement of the heart; (b) the posterior displacement of the cardiac apex; (c) partial rotation of the cardiac axis.

Analysis of MRI images was performed using a commercially available research software package (Mass Analysis, Leyden, The Netherlands). Ventricular volumes and mass were derived using the conventional method by the analysis of short axis cine images [17–19].

The end-diastolic and end-systolic transverse diameters (respectively DTD and STD) of both left and right ventricle have been measured as the maximal distance between the interventricular septum to the lateral wall in four-chamber view. The end-diastolic and end-systolic longitudinal diameters (DLD, SLD) have been measured as the distance between the atrioventricular valve and the ventricular apex for both ventricles in the same four-chamber view. All the diameters were indexed to the body surface area.

The systo-diastolic whole heart volume change (WHVC) was defined as the variation, expressed as a percentage, of the heart volumes between systole and diastole. WHVC was measured by tracing the contours around the entire heart in each image in the axial cine images in the end-systolic and end-diastolic phase. The contours included all cardiac structures within the pericardium (the atria, ventricles, and the roots of the aorta and pulmonary artery), excluding the great vessels (example in Figs. 1 and 2). The WHVC was then calculated as (end-diastolic heart volume-end-systolic)/end-diastolic volume, as a percentage (%).

The angle of clockwise-rotation of the heart was measured in the end-diastolic phase of the axial cine stack as the angle identified between the anterior–posterior line passing through the vertebral body and LV main axis (an example is shown in Fig. 3).

For the analysis of the intra-observer variability, Observer 1 has measured the same parameters (of both the LV and the RV) twice in the subjects. For the inter-observer analysis, Observer 2 has obtained measurements of the same subjects from a random sample previously analyzed by Observer 1.