Double-chambered left ventricle in an asymptomatic adult patient

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This article shows a case of double-chambered left ventricle in asymptomatic 60-year-old man. This rare congenital entity is usually diagnosed in the neonatal or paediatric age, and it is characterized by the subdivision of the left ventricle in two chambers by an abnormal septum or muscle band. Differential diagnosis includes two other congenital left ventricular (LV) outpouching entities, such as diverticulum and aneurysm. As shown in our case, double-chambered left ventricle exhibits mixed features. A comprehensive cardiovascular magnetic imaging study is useful in the distinction of these closely related congenital diseases and, therefore, important for the correct final diagnosis.

Keywords

Double-chambered left ventricle • Cardiovascular magnetic resonance

Double-chambered left ventricle is a particular rare congenital entity characterized by the presence of two ventricular cavities separated by an abnormal muscle band, and usually diagnosed during neonatal or paediatric age. In the current case report, we discuss an unusual presentation of double-chambered left ventricle in an asymptomatic adult patient, pointing out the key features for the differential diagnosis with two other similar congenital defects (diverticulum and aneurysm) by means of a comprehensive multimaging approach.

An asymptomatic 60-year-old-man underwent cardiac evaluation following the recent diagnosis of arterial hypertension. Physical examination was unremarkable. A 12-lead surface ECG showed sinus rhythm with left posterior hemiblock and right bundle branch block (Figure 1, left panel). At transthoracic echocardiography, the left ventricle had normal dimensions and function. However, the left ventricular (LV) lateral wall presented pronounced trabeculations interspersed by deep recesses (Figure 1, right panel). For this reason, the patient underwent cardiovascular magnetic resonance (CMR) (Figure 2). Balanced steady-state free-precession CMR confirmed prominent outpouching of the LV lateral wall separated from the main chamber (MC) by an abnormal muscle band, thus configuring an accessory chamber (AC) (Supplementary data online, Movies S1 and S2). The MC (end-diastolic volume 88 mL/m²) had normally contracting walls with preserved wall thickness and systolic function (ejection fraction 60%). On the contrary, the AC (end-diastolic volume 38 mL/m²) had a thinned and poorly contracting wall with decreased systolic function (ejection fraction 31%). On late post-contrast T1-weighted segmented gradient-echo inversion-recovery CMR, late gadolinium enhancement (LGE) was absent, thus excluding scarring of the AC and MC walls. Cardiac computed tomography confirmed the finding of the AC, and excluded critical stenoses of the main coronary arteries (Figure 3).

The sum of features of the present case is consistent with double-chambered left ventricle, a particularly rare congenital disease. Double-chambered left ventricle is usually diagnosed in the neonatal or paediatric age, and it is characterized by the subdivision of the left ventricle in two chambers by an abnormal septum or muscle band. Differential diagnosis includes two other congenital LV outpouching entities: diverticulum and aneurysm. The former is associated with congenital mid-line defects (i.e. Cantrell’s syndrome) and is characterized by a narrow connecting channel to the main ventricular cavity, along with a normally developed and contracting wall. In contrast, congenital aneurysm consists of a fibrotic saccular structure usually showing dyskinetic movement. As shown in our case, double-chambered left ventricle exhibits mixed features. Indeed, while the wide connection between the two chambers and the thinned and poorly contracting AC wall is common in congenital aneurysm, the absence of LGE on late post-contrast CMR indicates that the AC wall is constituted by normal myocardium as typically occurs in congenital diverticulum. A comprehensive...
**Figure 1** Twelve-lead electrocardiogram (left panel). Transthoracic echocardiography (right panel), parasternal (A) and four-chamber (C) views show the outpouching of the left ventricular lateral wall with deep recesses (asterisk). The abnormal muscle band (arrow) is interposed between the outpouched left ventricular wall and the main left ventricular cavity. However, colour-Doppler (B and D) disclosed a non-turbulent flow between the lateral wall recesses and the main left ventricular cavity.

**Figure 2** Cardiovascular magnetic resonance (images from A to I). End-diastolic (A, D and G), end-systolic (B, E and H) cine long-axis and short-axis images and the corresponding late post-contrast images (C, F and I) showing the prominent outpouching of the left ventricular lateral wall. Of note, the outpouched lateral wall was separated by the main chamber by an abnormal muscle band which delimited an accessory chamber made up of deep broad recesses (asterisk). On post-contrast images (C, F and I) there was no late gadolinium enhancement of the main chamber and accessory chamber walls.
CMR study is useful in the distinction of these closely related congenital diseases and, therefore, important for the correct final diagnosis.

**Supplementary data**

Supplementary data are available at European Journal of Echocardiography online.

**Conflict of interest:** none declared.

**References**