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Double trouble: The rare coexistence of double-chambered left ventricle with hypertrabeculation

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The double-chambered left ventricle (DCLV) is an extremely rare congenital defect with a challenging diagnosis due to complex and unobvious anatomy, which frequently calls for multimodality imaging, especially since alternative diagnoses, such as aneurysms, pseudoaneurysms, congenital diverticulum, or LV non-compaction cardiomyopathy, need to be carefully considered [1]. Although primarily asymptomatic and often discovered incidentally during unrelated diagnostics, this disease may have an unfavorable prognosis due to the risk of

malignant ventricular arrhythmias. Therefore, the appropriate diagnosis is of great clinical importance.

Although hypertrabeculation, with a prevalence of 0.56%, is not classified as a distinct entity [2], it may be associated with various cardiomyopathy phenotypes. However, it can also present as a normal variant or as a reversible adaptation to physiological conditions. The presence of isolated left ventricular hypertrabeculation has no established impact on prognosis or management in adults. Yet, the implications of this phenotype in cardiomyopathies, particularly in the context of coexisting DCLV, remain unclear [3].

A 20-year-old male was admitted for evaluation and potential implantable cardioverter-defibrillator implantation as primary prevention of sudden cardiac death, following a diagnosis of dilated cardiomyopathy of unknown etiology with reduced left ventricular ejection fraction 7 years earlier. At the age of 13, a chest X-ray was performed due to suspected cat scratch disease and lymphadenopathy, revealing significant cardiomegaly; an echocardiogram subsequently confirmed dilated cardiomyopathy with abnormal LV structure. Although the patient had no symptoms standard heart failure therapy, including angiotensin-converting enzyme inhibitors, beta-blockers, and mineralocorticoid receptor antagonists, was initiated. On the present admission patient was asymptomatic, with no laboratory abnormalities. Transthoracic and transesophageal echocardiography using 3D technology suggested DCLV with hypertrabeculation. The left ventricle consisted of a main (MLVC) and accessory chamber (ALVC), separated by a prominent muscular band without fully restricting communication. The ALVC, located at the base of the posterolateral wall, contained at least two pseudo-chords and resembled a large aneurysm with thin walls and excessive trabeculations as shown in Figure 1.

Magnetic resonance imaging confirmed the echocardiographic diagnosis (Figure 1, Supplementary material, *Figure S1* and *Video S1*), revealing an irregularly shaped, dilated LV with a double apex, segmental thinning up to 2 mm, isolated trabeculae extending into the LV cavity, and small regions of akinesia.

Ultimately, given the left ventricular ejection fraction above 35% and absence of symptoms, the patient was deemed ineligible for implantable cardioverter-defibrillator implantation and was discharged for continued outpatient careful monitoring.

Diagnosing DCLV *via* echocardiography is crucial but challenging [4], particularly in this case, where the band separating the MLVC and ALVC was not prominent. Additionally, identifying hypertrabeculation was complicated by difficulties in visualizing the apical segments of the posterior and lateral walls typically associated with this condition [5]. Notably, the hypertrabeculation observed in this case is atypical compared to prior literature, as it was

found exclusively in the ALVC. Comprehensive multimodality imaging, utilizing advanced echocardiography, including 3D technology and magnetic resonance imaging, enables early diagnosis of complex, rare, and potentially hazardous structural disorders within the myocardial tissue.

Supplementary material

Supplementary material is available at https://journals.viamedica.pl/polish_heart_journal.

Article information

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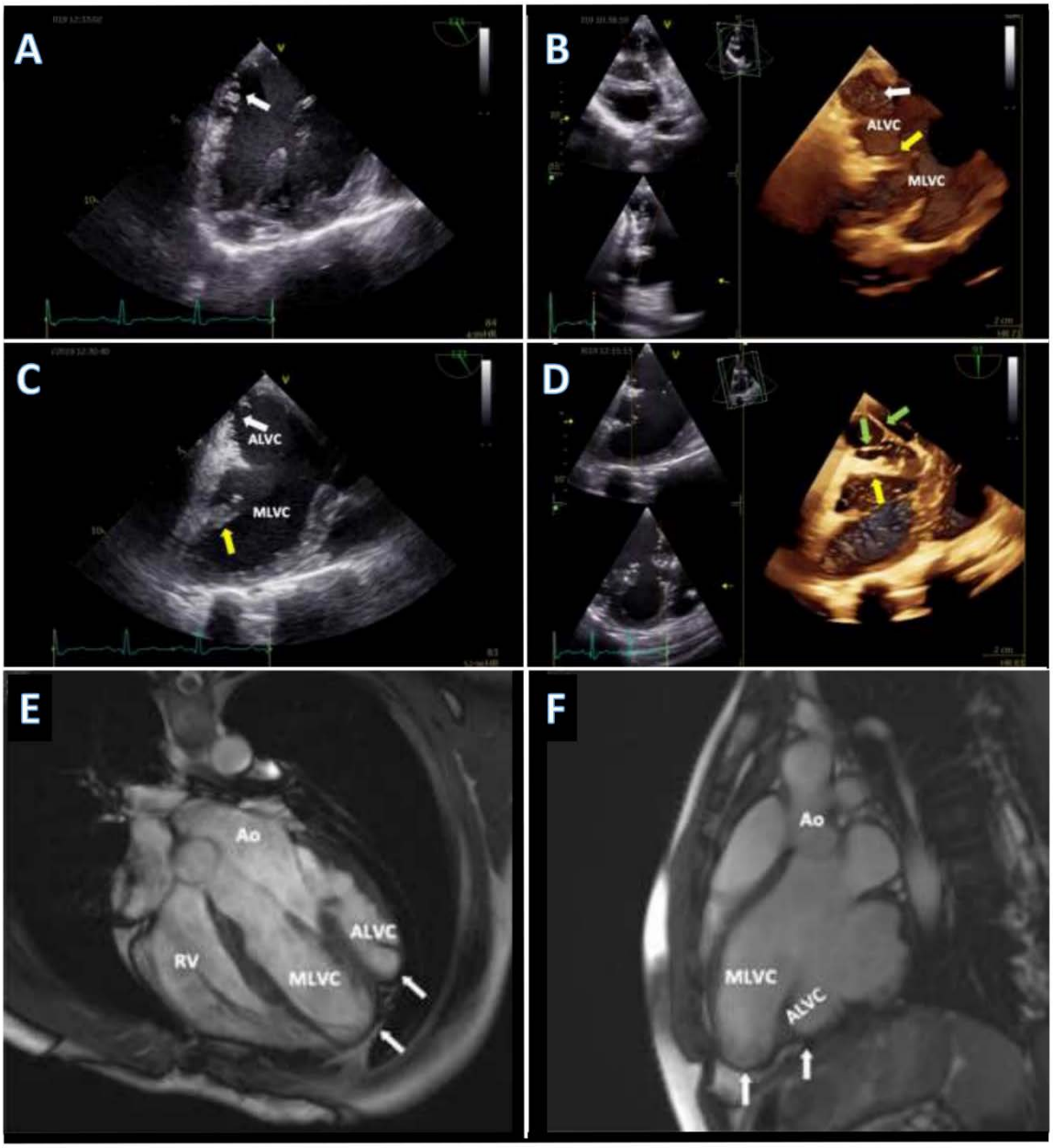

Figure 1. Transesophageal echocardiography demonstrates the ALVC and MLVC separated by a muscular band **(B, C, D**; yellow arrows), with the ALVC exhibiting hypertrabeculation (**A, B, C**; white arrows) and two pseudo-chords (**D**; green arrows). Cardiac magnetic resonance reveals an irregularly shaped LV with a double apex (**E, F**; white arrows)