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

Damian Kaufmann [1], Radosław Nowak[2], Jarosław Meyer-Szary[3], Karolina Dorniak[4,5], Michał Świątczak[1], Marcin Fijałkowski[6], Jadwiga Fijałkowska[5], Joanna Kwiatkowska[3], Ludmiła Daniłowicz-Szymanowicz[1]

[1]Department of Cardiology and Electrotherapy, Faculty of Medicine, Medical University of Gdansk, Gdańsk, Poland
[2]2nd Department of Cardiology and Electrotherapy, University Clinical Center, Gdańsk, Poland
[3]Department of Pediatric Cardiology and Congenital Heart Defects, Faculty of Medicine, Medical University of Gdansk, Gdańsk, Poland
[4]Department of Noninvasive Cardiac Diagnostics, Faculty of Medicine, Medical University of Gdansk, Gdańsk, Poland
[5]2nd Department of Radiology, Faculty of Health Sciences, Medical University of Gdansk, Gdańsk, Poland
[6]1st Department of Cardiology, Faculty of Medicine, Medical University of Gdansk, Gdańsk, Poland

**Correspondence to:**
Prof. Ludmiła Daniłowicz-Szymanowicz MD, PhD, Department of Cardiology and Electrotherapy, Medical University of Gdansk, Marii Skłodowskiej-Curie 3a, 80–210 Gdańsk, Poland, phone: +48 58 584 47 60, e-mail: ludwik@gumed.edu.pl



DOI: 10.33963/v.phj.103777

**Received:**
October 10, 2024

**Accepted:**
November 26, 2024

**Early publication date:**
December 9, 2024

A double-chambered left ventricle (DCLV) is an extremely rare congenital defect with a challenging diagnosis due to its 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, but 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 years, 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 the 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 2 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.

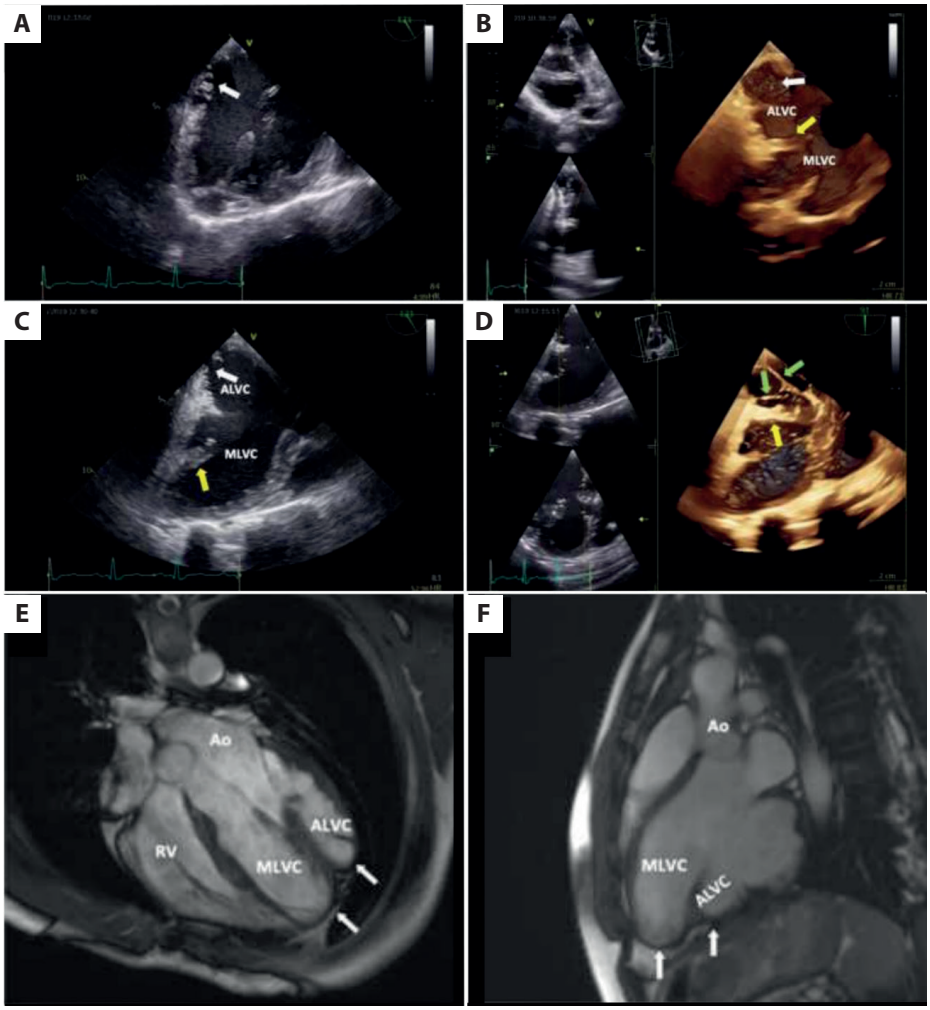


**Figure 1.** Transesophageal echocardiography demonstrates the ALVC and MLVC separated by a muscular band **(B, C, D**; yellow arrows), with the ALVC exhibiting hypertrabeculation (**A, B, C**; white arrows) and 2 pseudo-chords (**D**; green arrows). Cardiac magnetic resonance reveals an irregularly shaped LV with a double apex (**E, F**; white arrows)

Abbreviations: ALVC, accessory left ventricular chamber; Ao, aorta; MLVC, main left ventricular chamber; RV, right ventricle

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 because 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*

**Conflict of interest:** None declared.

**Funding:** None.



## REFERENCES

1. Nacif MS, Mello RAF, Lacerda Junior OO, et al. Double-chambered left ventricle in an adult: diagnosis by CMRI. Clinics (Sao Paulo). 2010; 65(12): 1393–1395, doi: 10.1590/s1807-59322010001200028, indexed in Pubmed: 21340234.
2. Arbelo E, Protonotarios A, Gimeno JR, et al. 2023 ESC Guidelines for the management of cardiomyopathies. Eur Heart J. 2023; 44(37): 3503–3626, doi: 10.1093/eurheartj/ehad194, indexed in Pubmed: 37622657.
3. Petersen SE, Jensen B, Aung N, et al. Excessive trabeculation of the left ventricle. JACC Cardiovasc Imaging. 2023; 16(3): 408–425, doi: 10.1016/j.jcmg.2022.12.026, indexed in Pubmed: 36764891.
4. Bao SF, Wu LJ, Zhang YQ, et al. Diagnosis of double-chambered left ventricle of superior-inferior type by echocardiography: A retrospective study of 9 subjects across two heart centers. Echocardiography. 2021; 38(2): 169–174, doi: 10.1111/echo.14939, indexed in Pubmed: 33368575.
5. Cuevas J, Ptaszynski R, Cigarrán H, et al. Left ventricular noncompaction cardiomyopathy: Recent advances. Kardiol Pol. 2022; 80(5): 529–539, doi: 10.33963/KP.a2022.0089, indexed in Pubmed: 35366003.