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Editorial Board Member of *World Journal of Clinical Cases*, Xin Ye, MD, Professor, Department of Oncology, The First Affiliated Hospital of Shandong First Medical University, Jinan 250014, Shandong Province, China. yexintaian2020@163.com

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CASE REPORT

# Double-chambered left ventricle with a thrombus in an asymptomatic patient: A case report

Narae Kim, In-Ho Yang, Hui-Jeong Hwang, Il-Suk Sohn

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**Narae Kim, In-Ho Yang,** Department of Medicine, Kyung Hee University, Seoul 02447, South Korea

**Narae Kim, In-Ho Yang, Hui-Jeong Hwang, Il-Suk Sohn,** Department of Cardiology, Kyung Hee University Hospital at Gangdong, Seoul 05278, South Korea

**Corresponding author:** In-Ho Yang, MD, Assistant Professor, Department of Cardiology, Kyung Hee University Hospital at Gangdong, 892 Dongnam-ro Gangdong-gu, Seoul 05278, South Korea. seventi4@hanmail.net

## Abstract

### *BACKGROUND*

Double-chambered left ventricle (DCLV) is an extremely rare congenital disease in which the left ventricle (LV) is divided by abnormal muscle tissue. Due to its rarity, there is a lack of data on the disease, including its diagnosis, treatment, and prognosis. Accordingly, we report a case in which DCLV was diagnosed and followed up.

### *CASE SUMMARY*

A 45-year-old man presented to our hospital due to abnormal findings on an electrocardiogram recorded during a health check. He had no specific cardiac symptoms, comorbidities or relevant past medical history. Echocardiography revealed that the LV was divided into two by muscle fibers. There were no findings of ischemia on coronary angiography and coronary computed tomography angiography performed to exclude differential diagnoses. After comprehensive analysis of the images, DCLV was diagnosed. As it seemed to be asymptomatic DCLV, we decided the patient was to be observed without administering any medication. However, follow-up echocardiography revealed a thrombus in the accessory chamber (AC). Anticoagulant medication was initiated, the thrombus resolved, and the patient is currently undergoing follow-up without any specific symptoms.

### *CONCLUSION*

Asymptomatic, uncomplicated DCLV was diagnosed through multimodal imaging; however, a thrombus in the AC occurred during the follow-up. The findings highlight that multimodal imaging is essential in diagnosing DCLV, and that anticoagulation is important in its management.

**Key Words:** Double-chambered left ventricle; Congenital heart disease; Left ventricular aneurysm; Echocardiography; Coronary computed tomography angiography; Contrast echocardiography; Case report



**Core Tip:** Double-chambered left ventricle (DCLV) is a rare congenital heart disease. Due to its rarity, the detailed data of the disease are not yet sufficient. We report the progress of a case of DCLV diagnosed incidentally. DCLV was diagnosed using multimodal imaging including echocardiography, coronary angiography, and coronary computed tomography angiography. After one year of follow-up, a thrombus occurred in the accessory chamber. The thrombus resolved after anticoagulation was initiated, and the patient is undergoing follow-up. Through this case, we demonstrate the necessity of multimodal imaging in the diagnosis of DCLV, and the importance of anticoagulation in the management.

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## INTRODUCTION

Double-chambered left ventricle (DCLV) is an extremely rare congenital heart disease in which the left ventricle (LV) is divided by abnormal muscle tissue[1-3]. Several studies report the prevalence of DCLV as around 0.04%–0.42%[1,3,4]. However, the exact prevalence is unknown, and few cases have been reported overall[1,3-5]. Due to the rarity of the disease, no conclusive data exist regarding its pathogenesis, diagnosis, treatment, and prognosis[1,3-5]. Most cases are diagnosed incidentally through an echocardiogram, coronary computed tomography angiography (CCTA), or cardiac magnetic resonance image (MRI) during diagnostic workup for other cardiovascular diseases[2,4]. Here, we report a case of a patient who was diagnosed with DCLV and is being observed under continued follow-up.

## CASE PRESENTATION

### *Chief complaints*

A 45-year-old man presented to the outpatient clinic at our hospital with abnormal electrocardiogram (ECG) findings.

### *History of present illness*

The patient underwent ECG as part of a regular health check, and had no cardiac symptoms such as chest pain or shortness of breath.

### *History of past illness*

He had no specific comorbidities and no history of trauma.

### *Personal and family history*

The patient had a smoking history of 20 pack-years and an alcohol consumption history of 3–4 bottles of soju per week. His younger brother was diagnosed with a myocardial bridge, and there was no other family history of cardiovascular disease.

### *Physical examination*

The patient had a height of 176 cm and weight of 73 kg, and his vital signs were stable, with a blood pressure of 121/74 mmHg and pulse of 100 beats/min. There were no other significant findings on physical examination.

### *Laboratory examinations*

Blood tests revealed no abnormalities, and cardiac markers were normal. The ECG showed T wave inversion in the anterior lead, and chest radiography findings were unremarkable.

### *Imaging examinations*

Echocardiography revealed that the LV was divided into two compartments by myocardial muscle fibers, along with tears in the apical lateral and inferior segments (Figure 1). There were no other significant abnormalities. Based on these findings, LV aneurysm dissection or LV pseudoaneurysm secondary to myocardial infarction (MI) were suspected.

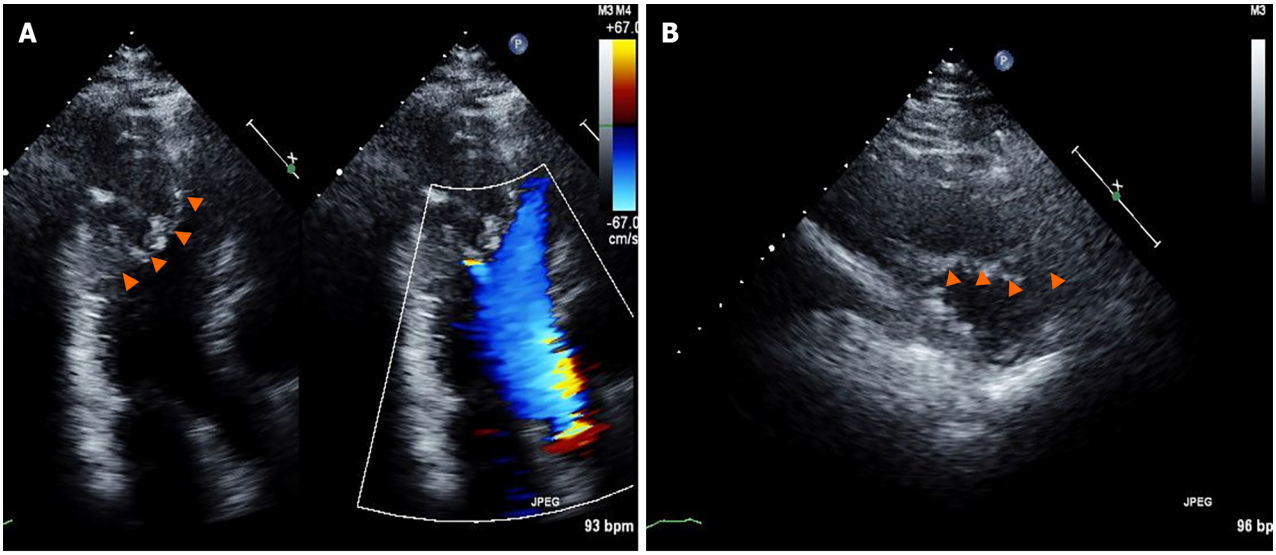


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**Figure 1 Echocardiography.** A: In the three-chamber view of echocardiography, the left ventricle (LV) is divided by an abnormal muscle bundle (orange arrows), along with tears in the LV apical lateral walls; B: The parasternal short-axis view of echocardiography shows the LV is separated by an abnormal muscle bundle (orange arrows) in the LV apical lateral and inferior walls.

### *Further diagnostic work-up*

Coronary angiography was performed to assess for coronary artery disease (CAD) such as MI. However, no significant luminal narrowing was observed. Although the findings regarding CAD did not appear, treatment was discussed with a thoracic surgeon in light of the possibility of aneurysmal dissection. After consulting with the surgeon, it was decided to prioritize a definitive diagnosis because surgical treatment in cases of asymptomatic aneurysm dissection is not urgent.

CCTA was performed for a more detailed evaluation of the anatomy and myocardial condition. On CCTA, it was shown that the LV was divided into two. A separated accessory chamber (AC) was connected to the LV by a narrow slit-like communication in the LV apical-mid posterolateral wall (Figure 2). No other abnormal findings, such as coronary artery anomaly, were seen, and no findings were suggestive of myocardial ischemia. Together, these findings indicated DCLV as a more likely diagnosis than LV pseudoaneurysm or aneurysm.

## FINAL DIAGNOSIS

Based on the multimodal imaging and patient's medical history, we diagnosed incidental DCLV.

## TREATMENT

We recommended cardiac MRI for a more detailed evaluation of the myocardium; however, the patient refused this further evaluation. The patient's vital signs were stable without any specific cardiac symptoms and showed normal heart function, so we decided not to perform the surgery. The patient was discharged and followed up at an outpatient clinic.

## OUTCOME AND FOLLOW-UP

The patient was followed up without any significant symptoms having occurred, and an echocardiography was performed 1 year later. Follow-up echocardiography showed no structural differences compared to the previous image; however, spontaneous echo contrast (SEC) with suspicion of a thrombus was visible in the AC (Figure 3). To check for the presence of a thrombus in the AC, a contrast echocardiography was obtained. On contrast echocardiography, a thrombus was suspected because the contrast was not transmitted along the apical inferior wall of the AC (Figure 3, Video). Additionally, although akinesia of the apical inferior segment was shown, the overall AC contracted during the systolic phase and dilated during the diastolic phase, consistent with a DCLV (Figure 3, Video). Anticoagulation was commenced using a direct oral anticoagulant to treat the thrombus in the AC. On echocardiography 6 wk later, SEC in the AC was persistent but the thrombus had resolved. We decided to continue anticoagulation prophylactically, and the patient has been attending follow-up as an outpatient without any development in his condition.

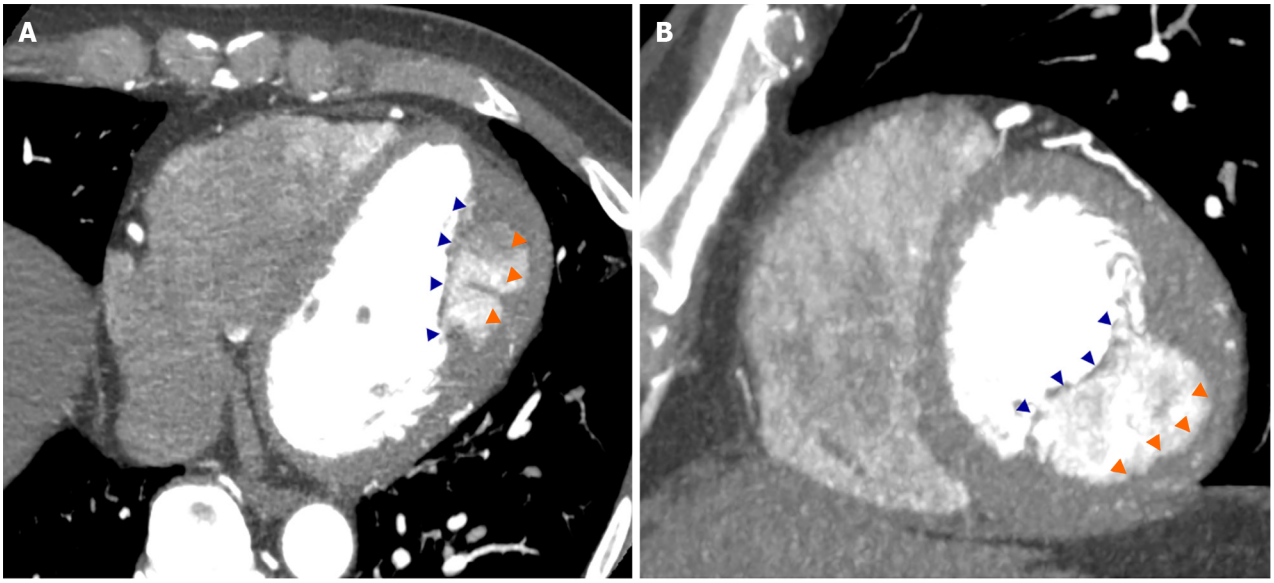


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**Figure 2 Coronary computed tomography angiography.** A: In the four-chamber view, two left ventricular (LV) chambers are divided at the lateral wall of the LV (blue arrows), and the accessory chamber is surrounded by normal myocardium (orange arrows); B: In the short-axis view, the LV chambers are surrounded by normal myocardium (orange arrows) and are separated at the posterolateral wall of the LV (blue arrows).

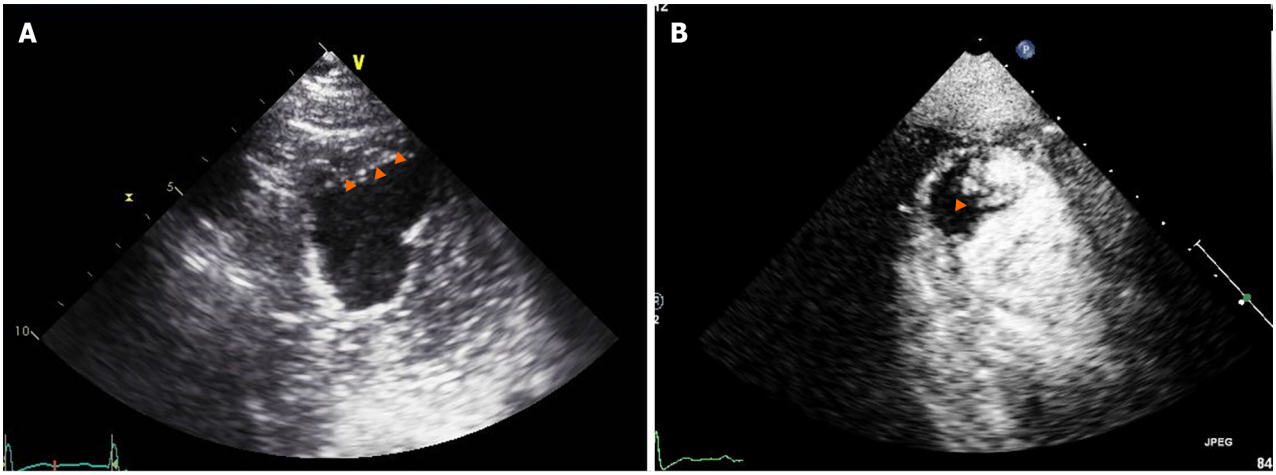


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**Figure 3 Follow-up echocardiography.** A: In the apical two-chamber view of the echocardiogram, spontaneous echo contrast with a probable thrombus (orange arrows) is shown in the accessory chamber. B: During contrast echocardiography, contrast does not penetrate along the apical inferior wall. The area (orange arrow) is probably a thrombus.

## DISCUSSION

DCLV is a very rare disease in which the LV is divided into two and is difficult to diagnose due to its rarity[6-8]. A diagnostic method to confirm DCLV has not been established[5,6]; however, DCLV should be differentiated from LV outpouching diseases, such as LV aneurysm or diverticulum[6,8]. As there are structural and etiological differences between these diseases, the definitive diagnosis is made using multimodal imaging techniques, such as echocardiography, coronary angiography, and cardiac MRI[6].

The difference between LV aneurysm/pseudoaneurysm and DCLV is that cases of aneurysm and pseudoaneurysm do not exhibit a full layer of ventricular wall[6]. For this reason, cases of aneurysm or pseudoaneurysm display dyskinetic wall motion abnormalities that expand in the systolic phase due to increased pressure[6]. In terms of etiology, aneurysm and pseudoaneurysm are mostly secondary to MI, whereas DCLV is innate[6,9]. In this case, partial wall motion abnormalities of the AC were suspected on echocardiography, making it difficult to diagnose. However, since there was no evidence of ischemia on coronary angiography and CCTA, the possibility of aneurysm or pseudoaneurysm is low in this case.

The difference between LV diverticulum and DCLV is that in LV diverticula, there is no abnormal septum or muscle fiber bundle dividing the LV[3,6]. Additionally, the former is often accompanied by congenital midline defects[6]. In this case, abnormal muscle fiber bundles dividing the LV were identified, and there was no congenital midline defect, which is more consistent with DCLV than LV diverticulum.

Gold standard treatment are not well established due to the rarity of DCLV[4,5]; however, conservative treatment or surgical correction is recommended based on the presence or absence of symptoms and complications[4,10]. The prognosis is also difficult to calculate[4]. Symptoms such as shortness of breath, chest discomfort, and palpitation may occur depending on the LV function and whether there is flow obstruction between the AC and the main chamber[5]. If complications such as heart failure or fatal arrhythmia are present, surgical treatment may be recommended[5,10,11]. If there are no symptoms and no complications, conservative treatment is recommended to prevent progression of the disease[5]. In this case, the SEC in the AC is prominent, meaning there is significant flow obstruction. Flow obstruction seemingly arose when blood flowed through the slit-like communication between the AC and the LV main chamber. Significant hemodynamic flow obstruction is thought to cause thrombosis[5]. Even in this case, thrombosis likely occurred in the AC due to hemodynamic flow obstruction. However, unlike in previously published cases, in this case, anti-coagulation was maintained, and progression of the disease was not observed.

## CONCLUSION

We report a case of DCLV in which the thrombus, having newly formed following DCLV diagnosis through multimodal imaging, was treated by anticoagulation and followed up. Through this case, we would like to highlight the importance of multimodal images in the diagnosis of DCLV and the importance of anticoagulation in its treatment.

## FOOTNOTES

**Author contributions:** Kim N contributed to manuscript writing and editing, and data collection; Hwang HJ and Sohn IS gave data; Yang IH contributed to conceptualization and supervision.

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**ORCID number:** Narae Kim 0000-0001-9759-9993; In-Ho Yang 0000-0002-2179-1767.

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