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# A Case of Double-Inlet Left Ventricle Reaching Adulthood Without Surgery

## Opere Edilmeden Erişkinliğe Ulaşmış Çift Girişli Sol Ventrikül Olgusu

**ABSTRACT**

Double-inlet left ventricle (DILV) is a rare congenital heart defect, also referred to as a single-ventricle defect. It has a complex structure in which blood from both atria flows into a single ventricle, accounting for approximately 1.5% of all congenital heart diseases. A 37-year-old female patient with no prior history of cardiac disease visited our outpatient clinic for routine cardiological evaluation. Transthoracic echocardiography (TTE) was performed after a 3/6 pansystolic murmur was heard at the mesocardiac focus and a 3/6 systolic ejection murmur at the pulmonary focus on cardiac auscultation. The patient's TTE revealed that the atrioventricular valves opened into one ventricular chamber in four-chamber apical imaging. There was no noticeable interventricular septum, while a rudimentary right ventricle and a ventricular septal defect (VSD) were observed. A gradient of 38 mmHg was measured in the pulmonary valve, and mild-to-moderate pulmonary stenosis was present. In the transesophageal echocardiography (TEE), a rudimentary interventricular septum and right ventricle were observed. Because the patient did not have any symptoms or cyanosis, she was informed about the potential need for occasional infective endocarditis prophylaxis and phlebotomy, and close medical follow-up was planned. A double-inlet left ventricle is known as a severe congenital heart anomaly that is typically diagnosed symptomatically in childhood and requires surgical intervention. However, very rarely, asymptomatic cases without surgical intervention have been reported in the literature to reach adulthood.

**Keywords:** Congenital heart diseases, double-inlet left ventricle, pulmonary stenosis, single ventricle, ventricular septal defect

**ÖZET**

Çift girişli sol ventrikül (DİLV), tek ventrikül defektleri olarak da bilinen, nadir görülen konjenital kalp defektlerindendir ve her iki atriyumdan çıkan kanın tek bir ventriküle aktığı karmaşık bir yapıya sahiptir ve tüm konjenital kalp hastalıklarının yaklaşık %1,5'ini oluşturur. Kalp hastalığı öyküsü olmayan 37 yaşındaki kadın hasta, rutin kardiyolojik değerlendirmeler için polikliniğimize başvurdu. Kardiyak oskültasyonda, mezokardiyak odakta 3/6 pansistolik ve pulmoner odakta 3/6 sistolik ejeksiyon üfürümü duyulması üzerine transtorasik ekokardiyografi (TTE) yapıldı. Hastanın TTE'sinde, dört odacıklı apikal görüntülemede atriyoventriküler kapakların tek bir ventriküler boşluğa açıldığı görüldü. Belirgin bir interventriküler septum görülmezken, rudimenter bir sağ ventrikül ve ventriküler septal defekt (VSD) izlendi. Pulmoner kapakta 38 mmHg'lik bir gradient vardı. Hafif-orta şiddette pulmoner stenoz mevcuttu. Daha sonra yapılan transözofageal ekokardiyografide (TEE) de rudimenter interventriküler septum ve sağ ventrikül gözlendi. Hastanın VSD'sinde şantın küçük düzeyde olması, pulmoner vasküler direncin normal sınırlarda bulunması ve belirgin semptom vermeden, siyanoz gelişmeden erişkin yaşa ulaşması da göz önüne alınarak ayrıca hastaya enfektif endokardit profilaksisi, ara ara flebotomi gerekebileceği anlatılarak yakın tıbbi takip kararı alındı. Çift girişli sol ventrikül, genellikle çocukluk çağında semptomatik olarak teşhis edilen ve cerrahi müdahale gerektiren ciddi bir konjenital kalp anomalisi olarak bilinir. Ancak literatürde, çok nadir de olsa cerrahi müdahale olmaksızın erişkinliğe ulaşan asemptomatik seyirli vakalar bildirilmiştir.

**Anahtar Kelimeler:** Konjenital kalp hastalıkları, çift girişli sol ventrikül, pulmoner stenoz, tek ventrikül, ventriküler septal defekt

**CASE REPORT**
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Double-inlet left ventricle (DILV) and double-outlet right ventricle (DORV) are rare congenital heart defects, also referred to as single-ventricle defects. They constitute approximately 1.5% of all congenital heart diseases.[1] In 70-75% of DILV cases, the main ventricle is in the form of a left ventricle (Figure 1). In most

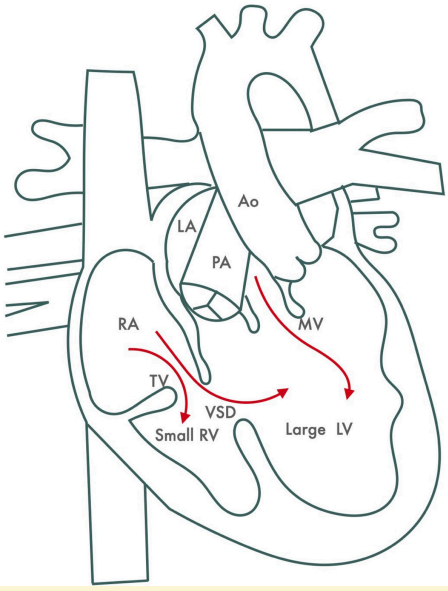


**Figure 1. Schematic illustration of double-inlet left ventricle.**

**Ao, Aorta; LA, Left atrium; LV, Left ventricle; MV, Mitral valve; PA, Pulmonary artery; RA, Right atrium; RV, Right ventricle; TV, Tricuspid valve; VSD, Ventricular septal defect.**

patients, the aorta arises from the hypoplastic right ventricle.[2] The prognosis of these cases is usually poor, and patients very rarely reach adulthood.[3] In general, diagnosis is made based on cyanotic symptoms in the early postpartum period, and surgical intervention is often required in the first years of life. Without surgical treatment, these cases usually result in death during infancy. This article presents a case of DILV in a patient who reached adulthood without diagnosis, complaint, or surgery.

## Case Report

The 37-year-old female patient with no history of cardiac disease visited our outpatient clinic for routine cardiological assessments. She did not report cardiovascular symptoms such as dyspnea, chest pain, palpitations, or syncope. Her medical history revealed only diabetes mellitus (DM), without other systemic or comorbid cardiac conditions. She had no history of cardiac medication use. The patient demonstrated normal physical and mental appearance and development. Her height was 162 cm, and her weight was 110 kg. The patient's functional capacity was classified as NYHA (New York Heart Association Functional Classification) class I. Her hemoglobin value was 16.0, and her hematocrit was 46.6%. Arterial blood gas analysis showed a partial oxygen pressure of 59.1 mmHg and a saturation of 90.3% (Table 1). Her blood pressure was 123/81 mmHg, heart rate 92 bpm, and respiratory rate 20 breaths per minute. She did not have cyanosis, and finger clubbing was not observed. On physical examination, a 3/6 pansystolic murmur was heard at the mesocardiac focus, and a 3/6 systolic ejection murmur was heard at the pulmonary focus on cardiac auscultation. Electrocardiography (ECG) showed a normal sinus rhythm. The ECG axis was normal, and the left ventricle was dominant. On telecardiography, the cardiothoracic ratio was normal.

## ABBREVIATIONS

| | |
|---|---|
| CT | Computed tomography |
| DILV | Double-inlet left ventricle |
| DM | Diabetes mellitus |
| NYHA | New York Heart Association Functional Classification |
| TEE | Transesophageal echocardiography |
| TTE | Transthoracic echocardiography |
| VSD | Ventricular septal defect |

**Table 1. The patient's laboratory values on admission**

| Parameter | Patient value | Reference range |
|---|---|---|
| Glucose (mg/dL) | 240 | 74–109 |
| Sodium (mmol/L) | 135 | 135–145 |
| Potassium (mmol/L) | 4.7 | 3.5–5.1 |
| Chloride (mmol/L) | 100 | 98–107 |
| Calcium (mg/dL) | 10 | 8.8–10.2 |
| Creatinine (mg/dL) | 0.7 | 0.7–1.2 |
| Blood urea nitrogen (mg/dL) | 18 | 8–23 |
| AST (U/L) | 27 | 10–50 |
| ALT (U/L) | 21 | < 41 |
| Albumin (g/dL) | 45 | 35–52 |
| TSH (mUI/mL) | 2.1 | 0.4–4.0 |
| pH | 7.40 | 7.35–7.45 |
| $pCO_2$ (mmHg) | 33 | 32–48 |
| $pO_2$ (mmHg) | 59 (low) | 83–108 |
| $sO_2$ (%) | 90 (low) | 95–99 |
| $HCO_3$ (mmol/L) | 22 | 22–28 |
| White blood cells ($10^9$/L) | 10.1 | 4.45–10.95 |
| Red blood cells ($10^{12}$/L) | 6.15 (high) | 4.54–6.11 |
| Hemoglobin (g/dL) | 16 (high) | 12–15.9 |
| Hematocrit (%) | 46.6 (high) | 36–46 |
| Platelet ($10^9$/L) | 364 | 162–367 |

ALT, Alanine aminotransferase; AST, Aspartate aminotransferase; $HCO_3$, Serum bicarbonate; $pCO_2$, Partial pressure of carbon dioxide; $pO_2$, Partial pressure of oxygen; $sO_2$, Oxygen saturation.

Transthoracic echocardiography revealed that the atrioventricular valves opened into a single ventricular chamber in four-chamber apical imaging. No noticeable interventricular septum was seen, while a rudimentary right ventricle and a ventricular septal defect (VSD) were observed (Video 1). Using the Simpson method, the ejection fraction of the common ventricle was calculated as 62%. There was a gradient of 38 mmHg across the pulmonary valve. Mild-to-moderate pulmonary stenosis was present. To observe the patient's cardiac anatomy in more detail, transesophageal echocardiography (TEE) was planned (Figure 2). In the TEE, the aortic valve had a tricuspid structure, the left atrial appendage was clear, and the interatrial septum was intact. A rudimentary interventricular septum and right ventricle were observed. The VSD was identified, and flow through the VSD was observed (Figure 3, Videos 2–6). A large portion of the VSD was covered by the septal leaflet of the tricuspid valve, with only a small

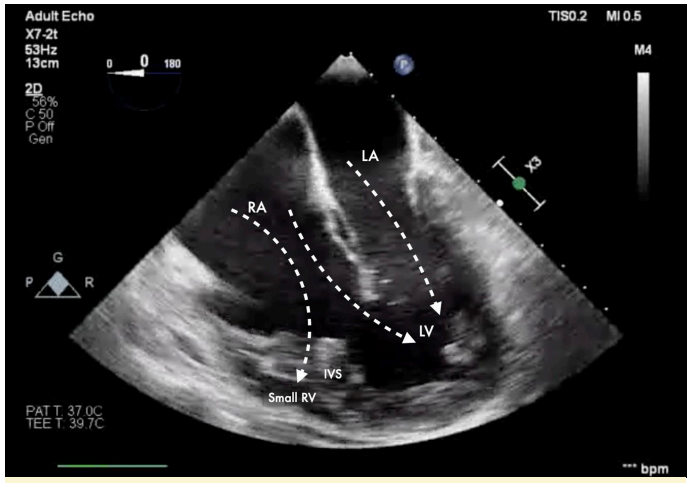


**Figure 2. Double-inlet left ventricle in mid-esophageal four-chamber view on transesophageal echocardiography (when the atrioventricular valves are open).**

**IVS, Interventricular septum; LA, Left atrium; LV, Left ventricle; RA, Right atrium; RV, Right ventricle.**

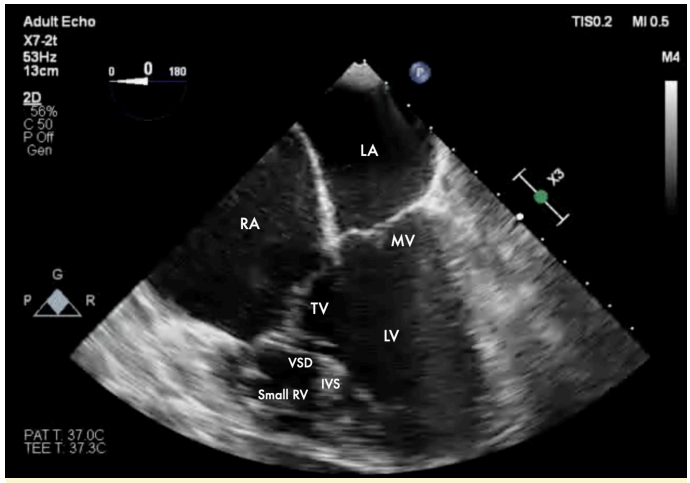


**Figure 3. Double-inlet left ventricle in mid-esophageal four-chamber view on transesophageal echocardiography.**

**IVS, Interventricular septum; LA, Left atrium; LV, Left ventricle; MV, Mitral valve; RA, Right atrium; RV, Right ventricle; TV, Tricuspid valve; VSD, Ventricular septal defect.**

segment showing right-to-left shunt flow. The right atrium was noticeably dilated. Cardiac catheterization was recommended. During cardiac catheterization, the pulmonary capillary wedge pressure was 15 mmHg, pulmonary artery pressure was 27/15/21 mmHg, right atrial pressure was 7 mmHg, and aortic pressure was 115/78 mmHg. Pulmonary vascular resistance was 1.9 Wood U, while systemic vascular resistance was 17.8 Wood U. Coronary angiography was also performed during the same session, and normal coronary arteries were observed. No anomalies such as coronary fistulae were detected. Computed tomography (CT) imaging was then performed. The CT results, compatible with echocardiography results, revealed a hypoplastic right ventricle, a rudimentary interventricular septum, and a VSD. No aortopulmonary collaterals were observed, and there was no transposition of the great arteries on CT. As the patient had no symptoms or cyanosis, she was counseled regarding the potential need for occasional infective endocarditis prophylaxis and phlebotomy, and close medical follow-up was recommended.

## Discussion

Single-ventricle anomalies may be frequently accompanied by pulmonary stenosis, pulmonary atresia, transposition of the great arteries, or aortic coarctation.[4] Moreover, in some DILV cases, the presence of anatomical and physiological factors that preserve hemodynamic balance may lead to an asymptomatic course of the disease and delayed diagnosis for years. A long-term asymptomatic course may be possible in the presence of pulmonary vascular resistance, atrioventricular valve structure, and minor comorbid anatomical variations.

In patients with single ventricles, anatomical features determine the prognosis, the severity of clinical findings, and the success of surgical methods.[5] Patients should be evaluated for the condition of the atrioventricular valves, major vascular anomalies, and ventricular type. Among single-ventricle defects, the left ventricular type is seen in 70–75% of cases, while the right ventricular type is seen in 10–15%. The ventricular type with both right and left ventricular features is known as the indeterminate type and is seen in 10–20% of cases. In follow-up studies based on anatomical features, a better prognosis was observed in the presence of DILV, transposition of the great arteries, and mild pulmonary stenosis.[6] In this combination, oxygenated blood arriving at the ventricle from the left atrium is directed toward the aorta due to pulmonary stenosis, while an amount of blood sufficient to prevent severe cyanosis but not cause volume overload passes to the pulmonary artery. The patient in our case fits the description of this combination.

Double inlet left ventricle is a rare congenital anomaly in which the atrioventricular valves open into a single left ventricle. The etiology of DILV is not clearly known; however, it is believed to involve genetic predisposition in addition to multiple factors. All patients with DILV have hypoxia, the severity of which is associated with the degree of shunting. In general, clinical symptoms are observed shortly after birth in these patients. The most common signs are dyspnea, tachycardia, cyanosis, and progressive cardiac insufficiency. In advanced cases, erythrocytosis and finger clubbing may develop.[7] These symptoms were not observed in our patient, who had reached adulthood, which is very rare.

Transposition of the great arteries is seen in 80–90% of patients, while pulmonary stenosis is present in 51%.[1] The long-term prognosis of DILV cases depends on the degree of pulmonary stenosis. In unoperated cases without pulmonary stenosis, a broad left-to-right shunt develops, leading to progressive cardiac insufficiency, fatal volume overload, and increased pulmonary blood flow. In the presence of pulmonary stenosis, pulmonary blood flow depends on the severity of the stenosis. Oliver et al.[3] reported that patients with L-transposition and pulmonary stenosis had the most favorable prognosis. In contrast, in a study of 83 unoperated patients with single-ventricle defects, Moodie et al.[8] argued that the severity of pulmonary stenosis did not have a significant effect on mortality.

The prognosis of patients with any form of single-ventricle defect and the onset of clinical symptoms largely depend on pulmonary arterial blood flow and pressure, which are influenced by the presence and severity of pulmonary stenosis and the level of pulmonary vascular resistance.[9,10] The degree of pulmonary blood flow and the severity of cyanosis determine the timing of surgical repair. In this case, mild-to-moderate pulmonary stenosis may have prevented pulmonary hypertension and maintained hemodynamic balance, allowing the patient to reach adulthood without symptoms.

The most important symptom in patients with single-ventricle defects is cyanosis observed after birth. Later in life, nonspecific complaints such as syncope, growth retardation, and exercise intolerance may also be observed. In patients with single-ventricle defects, ventricular systolic function supports both pulmonary and systemic circulation. This ventricular function, which is initially normal, may be impaired by pressure overload, volume overload, and comorbid pathologies.

Echocardiography, cardiac catheterization, and cardiac magnetic resonance imaging can be used in the diagnosis of DILV. In adults with complex congenital heart defects, echocardiography may be insufficient for functional assessments due to distorted ventricular geometry.

The most frequently encountered causes of mortality in these patients are arrhythmias, cardiac insufficiency, and sudden cardiac arrest. The survival rate of patients with single-ventricle defects within the first year of life is 30%.[11] The average life expectancy of non-operated patients ranges from 4 to 14 years.[8] This is why diagnosing and treating these patients at an early age is important.

In single-ventricle defects, step-by-step surgical treatment beginning in the neonatal period is the accepted form of treatment worldwide. It has been reported that the 10-year survival rate in patients suitable for the Fontan procedure reaches 81%.[12] While surgical treatment is unquestionable in neonatal cases, the situation differs for those detected in adulthood. According to Ammash et al.,[13] patients with well-developed pulmonary circulation may reach the sixth decade of life with good functional capacity. Deciding to keep these patients under follow-up is still difficult, as deterioration in their general condition during follow-up may render them unsuitable for the Fontan procedure. Cardiac transplantation is the last-resort treatment option for patients who have lost eligibility for the Fontan procedure.[14] The patient in our case was the fourth reported in adulthood in Türkiye. The other patients reached adulthood after refusing surgery after following diagnosis in childhood. Our patient did not show any symptoms until adulthood and therefore was not diagnosed. This suggests that she is a very rare case.

In the literature, the first report of a patient with a single-ventricle defect reaching adulthood was published by Goldberg et al.[10] Cases similar to ours, in which patients reached adulthood without surgery, have also been reported, although they are rare. While a few patients reaching the fifth to sixth decades of life have been described worldwide, before our case, only three such cases had been reported in Türkiye. Belgi et al.[15] presented a 21-year-old patient with a single-ventricle defect. Demir et al.[16] reported a 28-year-old male patient. Duran Karaduman et al.[17] described a 45-year-old female patient. In reports from outside Türkiye, Benelli et al.[18] reported a 63-year-old female patient diagnosed with congestive heart failure in advanced stages of life. Salame-Waxman et al.[19] reported a 28-year-old male patient diagnosed with pulmonary hypertension and aortic coarctation, who was followed without surgical intervention. David Gregg[20] described a male patient in his 50s with DILV who maintained stable vital signs and had an oxygen saturation of 80%.

The surgical option and the potential future consequences of not undergoing surgery were explained to our patient; however, she declined surgical intervention, stating that she currently had no significant symptoms. We decided to schedule regular follow-ups, as her VSD had only a small shunt, her pulmonary vascular resistance was within normal limits, and she had reached adulthood without noticeable symptoms or cyanosis. The patient was also informed that she might require infective endocarditis prophylaxis and phlebotomy. Diuretic treatment was not necessary.

This case highlights the protective role of pulmonary stenosis and the rare possibility of reaching adulthood without surgery, while underlining the importance of close follow-up and prophylactic measures.

## Conclusion

A double-inlet left ventricle is a severe congenital heart disease that is rarely seen in adulthood, is usually diagnosed in childhood, and requires surgical intervention. Our case demonstrated that double-inlet left ventricle has a broad clinical spectrum: some patients may have a long-term asymptomatic course, and in rare instances, the condition may first be diagnosed in adulthood. The importance of routine cardiological assessments, physical examination, and meticulous imaging techniques in diagnosing congenital heart defects with a silent course was also highlighted. Such cases diagnosed in adulthood serve as a reminder of the need to consider congenital heart disease not only in the pediatric population but also in adults.

**Ethics Committee Approval:** This is a single case report, and therefore ethics committee approval was not required in accordance with institutional policies.

**Informed Consent:** Informed consent was obtained from the patient.

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**Video 1.** Double-inlet left ventricle in apical four-chamber view on transthoracic echocardiography.

**Video 2.** Double-inlet left ventricle in mid-esophageal four-chamber view on transesophageal echocardiography.

**Video 3.** Double-inlet left ventricle in mid-esophageal four-chamber view on transesophageal echocardiography (180º).

**Video 4.** Double-inlet left ventricle on transesophageal echocardiography.

**Video 5.** Double-inlet left ventricle on transesophageal echocardiography (slow motion).

**Video 6.** Double-inlet left ventricle in mid-esophageal four-chamber view on transesophageal echocardiography.

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