# Mitral valve with a single leaflet

## Tek yaprakçıklı mitral kapak

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***Summary–*** Congenital anomalies of the mitral valve apparatus are rare. Of such cases, congenital mitral stenosis, atresia, accessory valvular tissue, and cleft mitral valve are more common. Descriptions of unileaflet mitral valves (either partial or complete leaflet agenesis/hypoplasia) are extremely rare and largely limited to a few case reports. We report herein a 45-year-old nulliparous woman presenting to our outpatient clinic with chest discomfort and dyspnea on minimal exertion. Mitral valve with a single leaflet leading to a significant obstruction in left ventricular outflow was seen on transthoracic echocardiography and confirmed with transesophageal two- (2D) and three-dimensional (3D) echocardiography. Metoprolol was prescribed to relieve obstruction and symptoms.

***Özet–*** Mitral kapağın doğumsal bozuklukları nadirdir. Bunlardan doğumsal mitral darlığı, atrezisi, aksesuvar kapak dokusu ve kleft mitral kapak daha sıktır. Tek yaprakçıklı mitral kapak (kısmi veya tam yaprakçık agenezisi/hipoplazisi) olguları ile ilgili bildiriler çok az olup ve bir kaç olgu ile sınırlıdır. Bu yazıda, hafif eforla nefes darlığı ve göğüste huzursuzluk şikayetleri ile polikliniğimize başvuran 45 yaşında hiç doğum yapmamış bir kadın hasta sunuldu. Transtorasik ekokardiyografide sol ventrikül çıkış yolunda ciddi obstrüksiyona sebep olan tek yaprakçıklı mitral kapak tespit edildi. Tanı iki boyutlu ve üç boyutlu transözofajiyal ekokardiyografi ile doğrulandı. Obstrüksiyonu ve semptomları hafifletmek için meteprolol başlandı.

Congenital anomalies of the mitral valve apparatus are rare. Of such cases, congenital mitral stenosis, atresia, accessory valvular tissue, and cleft mitral valve are more common. Descriptions of unileaflet mitral valves (either partial or complete leaflet agenesis/hypoplasia) are extremely rare and largely limited to a few case reports.

We report herein a 45-year-old nulliparous woman presenting to our outpatient clinic with chest discomfort and dyspnea on minimal exertion.

**Abbreviations:**

| | |
|---|---|
| *2D* | *Two-dimensional* |
| *2DE* | *Two-dimensional echocardiography* |
| *3D* | *Three-dimensional* |
| *3DE* | *Three-dimensional echocardiography* |
| *TTE* | *Transthoracic echocardiography* |

## CASE REPORT

A 45-year-old woman presented to our outpatient clinic with dyspnea on exertion. She had been operated for strabismus and ptosis three years earlier. The physical examination revealed pectus carinatum and hypertelorism. The patient had no child due to male factor infertility. She is the single surviving member of her family. There was no history of congenital heart disease in her family. She was screened for chromosomal and congenital abnormalities with genetic testing, and the results were negative. In addition, she was consulted to the Department of Obstetrics and Gynecology, which reported that the patient had functioning ovaries and a positive history of regular menstrual cycles.

The ECG demonstrated sinus rhythm and loss of R-progression on chest leads. A systolic murmur was heard on auscultation. On transthoracic echocardiography (TTE), the posterior mitral leaflet was not visu-

*Received:* May 14, 2013 *Accepted:* July 30, 2013
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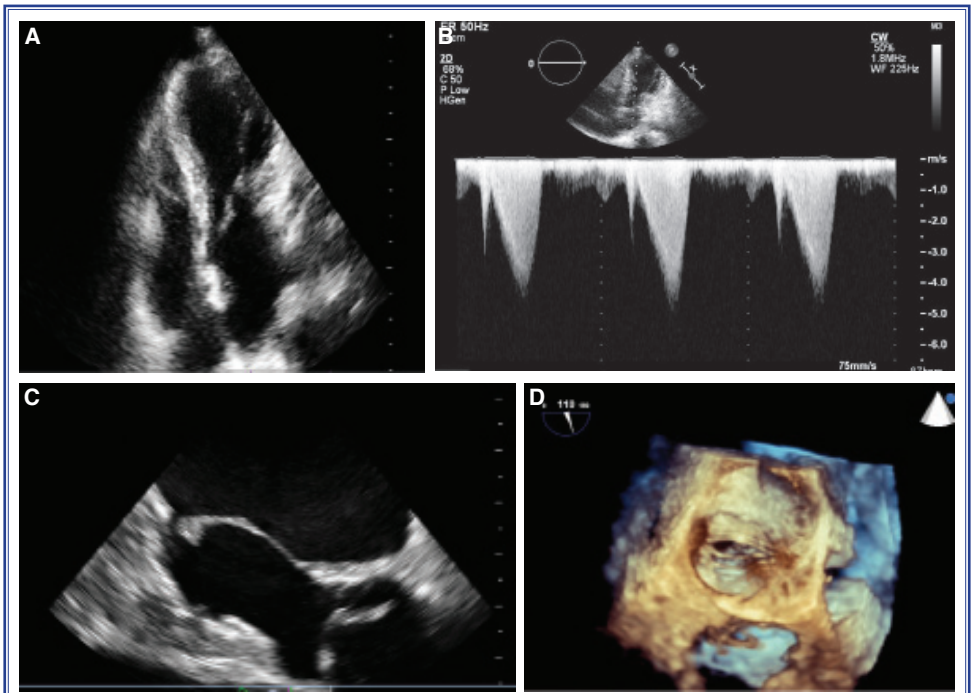


**Figure 1.** **(A)** Transthoracic apical four-chamber view showing the protrusion from the left ventricular free wall, which functions like a posterior leaflet. **(B)** There was a significant gradient in the left ventricular outflow tract at rest. **(C)** Transesophageal image showing the absence of the posterior leaflet. **(D)** Three-dimensional transesophageal image with left atrial perspective showing the absence of the posterior leaflet.

alized. A left ventricular free wall had replaced it and was coapting with the anterior leaflet (Fig. 1a; Video 1*). The anterior leaflet and chordal structures were elongated. Systolic anterior motions of the mitral leaflet and posterior annulus were present. Peak and mean gradients of 100 and 50 mmHg, respectively, were measured in the left ventricular outflow tract at rest (Fig. 1b). Trace mitral and aortic regurgitations were also present. On two-dimensional (2D) and real-time three-dimensional (3D) transesophageal echocardiography, no leaflet was detected at the localization of the posterior leaflet, as in the TTE (Fig. 1c, d; Videos 2, 3*). The free wall of the left ventricle that had replaced the posterior leaflet was found to coapt with the anterior mitral leaflet. The chordal structures were seen to extend from the papillary muscle to the ventricular wall, which had taken the form and function of the missing posterior leaflet. Metoprolol (50 mg once daily) was initiated to relieve obstruction in the left ventricular outflow tract and symptoms.

## DISCUSSION

Congenital anomalies of the mitral valve apparatus are seen very infrequently. Of such cases, congenital mitral stenosis, atresia, accessory valvular tissue, and cleft mitral valve are more common.

Reports of functionally unileaflet mitral valves (either partial or complete leaflet agenesis/hypoplasia) are extremely rare.[1-5] It was reported that unileaflet mitral valve can show familial clustering.[6] In the most severe form (complete leaflet absence), cases are usually considered to be incompatible with life beyond the neonatal period. Asymptomatic patients, however, do exist, and a prevalence of 1:8800 has been documented previously in preselected patient cohorts.[5]

Three-dimensional echocardiography (3DE) is a simple, quick and relatively well-established imaging modality that offers several advantages over traditional two-dimensional echocardiography (2DE) in the examination of the anatomy and function of each of the individual components of the mitral valve apparatus, including the annulus, leaflets, chordae, and papillary muscles. The 3D evaluation may be easily integrated into the standard 2D examination in the assessment of the qualitative morphology of the mitral

valve.[7-10] Its capability to image and represent the mitral valve apparatus in its intrinsic 3D nature has improved our understanding of the normal and abnormal mitral valve. A substantial body of literature evidences the advantages of 3DE over conventional 2DE, underlining the pros during the diagnostic phase as well as during interventional and surgical procedures.

We report herein a case of unileaflet mitral valve, which is infrequently encountered, together with the better delineation of the mitral valve apparatus with 3DE. Long-term monitoring with serial echocardiography seems appropriate.

***Conflict-of-interest issues regarding the authorship or article: None declared.***

****Supplementary video files associated with this article can be found in the online version of the journal.***

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*Key words:* Echocardiography, three-dimensional; heart defects, congenital/ultrasonography; mitral valve/abnormalities; unileaflet mitral.

*Anahtar sözcükler:* Ekokardiyografi, üç-boyutlu; kalp defekti, doğuştan/ultrasonografi; mitral kapak/anomali; tek yaprakçıklı mitral kapak.