

# Double Orifice Mitral Valve and Spontaneous Echo Contrast in the Descending Aorta

## *Desandan Aortada Eko Kontrast ve Çift Orifis'li Mitral Kapak*

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### Introduction

Double orifice mitral valve (DOMV) is a rare congenital anomaly of the subvalvar mitral valve apparatus (chordae tendinae and papillary muscles) consisting of an accessory bridge of fibrous tissue, which partially or completely divides the mitral valve into two orifices (1). Spontaneous echo contrast (SEC) in aorta is more commonly seen in patients with severely reduced cardiac output, in the vicinity of dilated or aneurysmal aortas or within the false lumen created by aortic dissection. However, it has also been noted in the absence of structural heart disease and in the presence of normal aortic dimensions (2).

To the best of our knowledge the presence of these two conditions together in the same patient has not been reported in the literature. In this report, we describe an 80-year-old male patient with DOMV, SEC in the descending aorta lumen without aortic dissection which were diagnosed by transthoracic and transesophageal echocardiography.

### Case Report

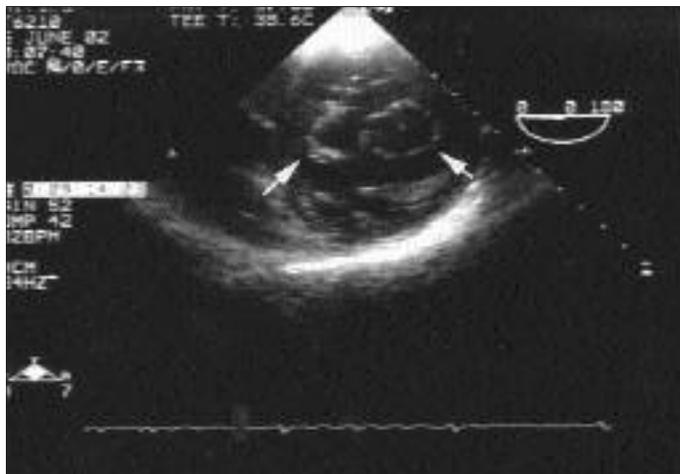
A 80-year-old man was admitted to the hospital with dyspnea and angina on effort for eight months. He had no familial history of heart disease and coronary risk factors. In his physical examination, blood pressure was 140/70 mmHg, his pulse was 100-110 bpm and irregular. He had a grade 3/6 mesocardiac and apical pansystolic murmur on cardiac auscultation. The other system examinations were normal. Electrocardiogram showed atrial fibrillation with signs of left ventricular hypertrophy and negative T waves in V1 to V5 leads. Chest X-ray revealed mildly increased cardiothoracic index and normal pulmonary vasculature. Transthoracic echocardiography in the short-axis view at mitral valve level and transesophageal echocardiography in transgastric views (Fig. 1) revealed two orifices in the mitral valve with nearly equalized size. Color Doppler echocardiography showed severe mitral regurgitation from both orifices in the api-

cal four-chamber view. Left ventricular (LV) ejection fraction was 55%, LV end-systolic diameter- 42 mm, LV end-diastolic diameter- 58 mm and pulmonary artery systolic pressure was 55 mmHg. Mitral stenosis was not detected. Mild aortic regurgitation was shown in apical five chamber views. Transesophageal echocardiography revealed double mitral orifices, severe mitral regurgitation and each orifice had its own subvalvular apparatus (Fig. 2). No signs of aortic dissection were determined. Ascending and descending aortic diameters were quite large; 39 and 31 mm, respectively. Severe SEC in the descending aorta was also observed in the transesophageal echocardiographic views (Fig. 3). Aortic wall calcification but not complex plaque was another finding in the descending aorta. Thoracic computerized tomography scan showed large ascending aorta dilation (40 mm). Aortic dissection or aneurysm was not observed. The patient refused further evaluation and therapy.

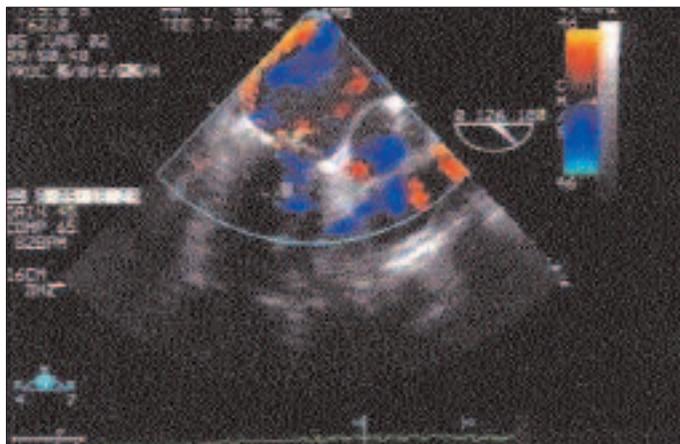
### Discussion

Since the first publication by Greenfield (1), approximately 200 cases of DOMV have been recognised incidentally at necropsy (3), surgery (4, 5) or by echocardiography (5). Double orifice mitral valve may be associated with a variety of other cardiac anomalies such as coarctation of aorta, subaortic ring, bicuspid aortic valve, patent ductus arteriosus, pulmonary stenosis, atrioventricular septal defects, truncus arteriosus and hypoplastic left heart syndrome (6-8). Functionally, the mitral leaflets are essentially normal in most cases (3, 4) but they can be regurgitant (3) as presented in our case or stenotic (3). Cases with equal orifices similar to our patients are less frequent (3). Two-dimensional echocardiography is a useful method for the diagnosis of double orifice mitral valve. Two separate holes in the mitral valve can be identified in parasternal short-axis view and DOMV may be more detectable using transesophageal echocardiography in short-axis transgastric views of two mitral orifices (Fig. 1). Especially color Doppler echocardiography provides adequate anatomical and functional assessment of DOMV (7).

The clinical implication of DOMV depends on the concomitant pathology. In the absence of other anomalies it usually has no hemodynamic consequences. However, our case had symptoms such as dyspnea on effort as a result of severe mitral regurgitation.



**Figure 1.** Transgastric short-axis view of transesophageal echocardiography showing two separate orifices of mitral valve nearly equalized in size



**Figure 2.** Color Doppler image of transesophageal echocardiography showing severe mitral regurgitation from two separate orifices of mitral valve



**Figure 3.** Severe spontaneous echo contrast detected in the descending aorta with transesophageal echocardiographic examination

The surgical correction of DOMV depends on the patient's condition. Three strategies have been adopted in the correction of DOMV: major valve repair, cleft suture and valve replacement (3). When DOMV occurs in a functional and asymptomatic valve, it should not be touched. Severe mitral regurgitation similar to our patient could undergo valve repair. Because superiority of valve repair to valve replacement is generally approved, we should attempt to perform valve repair for mitral regurgitation with DOMV.

Spontaneous echo contrast in the descending aorta in patients without aortic dissection was found to be associated with older age, male gender, larger diameters of ascending and descending aorta, aortic wall calcification and complex plaque in the descending aorta, and left ventricular dysfunction (9). Reported frequency of the aortic SEC varies between 0.8 and 21% in the transesophageal echocardiography series (10). In this case, the cause of SEC in the descending aorta may be low cardiac output due to severe mitral regurgitation. Another explanation may be also large diameter of aorta and aortic wall calcification.

Our case is unusual for being the oldest person with DOMV in the literature. Presence of SEC in the descending aorta and DOMV also have never been reported together in the same patient.

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