Isolated double orifice mitral valve and spontaneous echo contrast without dissection in the descending aorta

İzole çift orifisli mitral kapak ve desandan aortada disseksiyon olmaksızın spontan EKO kontrast

Dear Editor,

Double orifice mitral valve (DOMV) was firstly described by Greenfield in 1876 (1). This anomaly is a rare congenital malformation reported in 1% of postmortem cases of congenital heart diseases (2).

The embryological formation of DOMV has not been completely understood, yet. The abnormality of invagination of the atrioventricular sulcus and undermining of ventricular myocardium can lead to such a defect (3). It has been suggested that DOMV is the result of abnormal differentiation of mesenchymal endocardial cushion tissue into chordae instead of leaflet tissue, thereby producing the accessory orifice (2).

This abnormality may rarely present as an isolated lesion, but, most commonly, it is associated with other cardiac anomalies such as atrioventricular septal defects which are the most commonly associated lesions, bicuspid aortic valve, coarctation of the aorta, subaortic ring, patent ductus arteriosus, pulmonary stenosis and Ebstein's anomaly (2,4-7). In the series of Zalzstein et al. (5) only 3 of the 46 patients were found to have an isolated DOMV. Therefore, it must be routinely assessed in patients especially with atrioventricular septal defects.

The clinical presentation of DOMV is variable and depends mainly on the associated, more complex cardiac lesions. There are no specific clinical signs to suggest DOMV, both electrocardiogram and chest X-ray are usually normal. Echocardiography represents the diagnostic method of choice to detect the malformation. Two-dimensional echocardiography with the Doppler technique is undoubtedly the best method; the appearance of 2 separate orifices in parasternal short-axis view is diagnostic of DOMV. With a parasternal short-axis approach, differential diagnosis should be made against the cleft anterior mitral leaflet and diastolic pattern of anterior mitral leaflet in severe aortic insufficiency. The superiority of transesophageal echocardiography over transthoracic echocardiography in patients with DOMV has been previously reported. The key to the correct anatomic diagnosis of this rare malformation is the deep transgastric view (8).

Trowitzsch et al. (9) recognized 3 different types of DOMV using 2-dimensional echocardiography.

1- Complete bridge type: Two separate, complete orifices are visible from the leaflet edges all the way through the valve ring. Both orifices are circular and almost equal in size with the appearance of spectacles.

2- Incomplete bridge type: Anterior and posterior leaflets are connected only at the leaflet edge, and only at this level the double-orifice is visible. At mid- and basal level the mitral valve appears normal.

3- Hole type: A single orifice is present at the leaflet tips, and additional smaller orifice is visualized in one of the two commissures of the mitral valve only at the mid-leaflet level and disappearing on scanning toward the apex or base.

These three types can be distinguished by sweeping the transducer in cross-sectional view from the apex toward the base of the heart. Both orifices can be seen throughout the scan in the complete bridge type while in the incomplete bridge type the two orifices can be seen only at the level of the papillary muscles. In the hole type, the second orifice is seen at about midleaflet level. The apical four-chamber view may be particularly useful in patients with hole-type defects because in these cases the two orifices do not lie in the same plane and thus they cannot be recorded in short-axis view.

In most cases, DOMV causes no hemodynamic effects, and can be detected accidentally by echocardiography. Sometimes it is regurgitant, and stenosis is rarely present. On the basis of the hemodynamic effects of the mitral valve, 4 groups of patients can be identified: 1- those with no functional importance, 2- those with mitral insufficiency only, 3- those with mitral stenosis only, and 4- those with mitral stenosis and insufficiency. Mitral insufficiency is the most common functional abnormality, and mitral stenosis, either alone or with insufficiency, is rare. The presence of atrial septal defects in many children can lead to the underestimation of the frequency and/or severity of mitral stenosis, which would be unmasked after atrial septal defect closure (5). The color Doppler method is useful in cases of difficult standard echocardiographic images allowing to identify precisely the different flows with functional and hemodynamic information of the abnormality.

When a diagnosis of double-orifice mitral valve is suspected, echocardiographic study must not be limited to valve leaflets but should involve the tensor apparatus that always appears anomalous. The key to the diagnostic and surgical understanding of the double-orifice mitral valve is the underlying tensor apparatus. This approach serves not only to distinguish this congenital malformation among the other mitral leaflet abnormalities, but also to describe the relationship between the valve leaflets and subvalvular structures and to better assess the hemodynamic consequences by the Doppler technique. The papillary muscles are commonly located in normal way. Typically the chordae from the lateral (anterior) orifice is attached only to anterolateral papillary muscle, whereas the chordae of the medial (posterior) orifice is attached to the posteromedial papillary muscle creating a kind of double parachute valve (10).

Double-orifice mitral valve almost always consisted of abnormal holes in essentially normal leaflets, rather than of abnormal fibrous bridges or adhesions between normal leaflets. Since these fibrous "bridges" between the smaller accessory orifice and

the larger main orifice are composed of mitral leaflet tissue and chordae, not fibrous adhesions, these bridges should not be transected surgically, to avoid iatrogenic mitral regurgitation.

The surgical intervention is not necessary in patients with isolated DOMV unless they also suffer from mitral stenosis and/or mitral regurgitation. When the surgery is needed, superiority of valve repair to valve replacement is generally approved. However, mitral valve replacement has been also reported for patients with DOMV (11).

In the article published in the past issue of the Anatolian Journal of Cardiology, a 80-year-old male patient with isolated some was reported (12). First of the features of the present case is rarity of isolated DOMV as an anomaly, and second is its failure to be diagnosed until the age of 80 despite its co-existence with significant mitral insufficiency. Heyse et al. (13) have recently reported the case of a 79-year-old woman with mitral insufficiency and isolated DOMV (13). Whereas, additionally, spontaneous echo contrast (SEC) in the descending aorta, which has not been previously reported in patients with DOMV, was also demonstrated in the current patient.

The visualization of SEC is a common phenomenon in patients undergoing transesophageal echocardiographic studies. Prevalence of SEC in the descending aorta in the absence of dissection has been reported as 4.5% (14). Its pathophysiology is not well understood, but it has been related to the presence of a low flow state in the cardiac chambers. Although SEC in the descending aorta has also been noted in the absence of structural heart disease and in the presence of normal aortic dimensions (15-18), several factors that have been previously shown to be related to SEC in the descending aorta (14,17) such as male gender, older age, atrial arrhythmia, dilatation of ascending and descending aorta and calcification on the aortic wall were present in this patient. The low cardiac output due to the significant mitral insufficiency may be a cause of SEC in the descending aorta. Furthermore, laboratory indexes of thrombotic status or platelet activity might also be taken into account as a cause of SEC.

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Author's Reply

Dear Editor

We would like to express our gratitude to the author of the letter to the Editor, for the contributions regarding our case report published in the September 2004 issue of the journal.

Sincerely

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