

## ORJİNAL GÖRÜNTÜLER ORIGINAL IMAGES

### Isolated Double Orifice Mitral Valve

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Double-orifice mitral valve is a rare congenital malformation. Since first described by Greenfield in 1876, approximately 230 cases have been reported. (1). The isolated occurrence of this anomaly is exceptional and, more often, is encountered in association with other congenital cardiac abnormalities such as atrioventricular septal defects, bicuspid aortic valve and coarctation of aorta. The hemodynamic effects of this malformation is variable and although valve functions are frequently normal regurgitation or stenosis may be present. Before echocardiography was widely used, detection of this lesion was incidental at autopsy or during surgical correction of associated cardiovascular defects. Echocardiography has allowed a noninvasive detection of this abnormality.

We describe a case of double-orifice mitral valve diagnosed by echocardiography without any associated abnormality;

A 34-year old man was referred for cardiology examination because of chest pain that was present for 3 years. The pain, burning, stabbing in character, was unrelated to effort and it was radiating toward right arm and backward. Physical examination was normal, except for a soft, short grade 1/6 systolic murmur heard at meso-cardiac area. Chest X-ray, complete blood count and biochemical analysis were normal. Electrocardiogram showed sinus rhythm and incomplete right bundle branch block. On treadmill exercise (modified Bruce protocol) he achieved 9 MET's without any symptoms and associated ST-segment depression.

Transthoracic echocardiography showed a V-shaped mitral valve in apical two chamber and subxiphoid views. Cardiac chambers were normal in size. The transesophageal echocardiography clearly showed that there were two separate valve orifices with separate leaflet structures, two cusps with a single horizontal coaptation line attached to a raphe presenting as V-shaped ("seagull wing") mitral valve (Figure 1). The orifices were almost equal in size. There were no evidence of additional associated pathologies, valvular regurgitation or stenosis.

Transgastric short axis view showed that there were four papillary muscles, each leaflet having its own chordae and papillary muscle (Figure 2).

From echocardiographic point of view three types of double-orifice mitral valve have been described; 1. Complete bridge type: two separate, complete orifices being circular in shape and almost equal in size; the absence of associated cardiac malformation and hemodynamic abnormality is more frequent in patients with double mitral orifices of equal size (2), 2. Incomplete bridge type: anterior and posterior leaflets are connected



Figure 1: "Seagull wing" mitral valve appearance at transesophageal echocardiography.



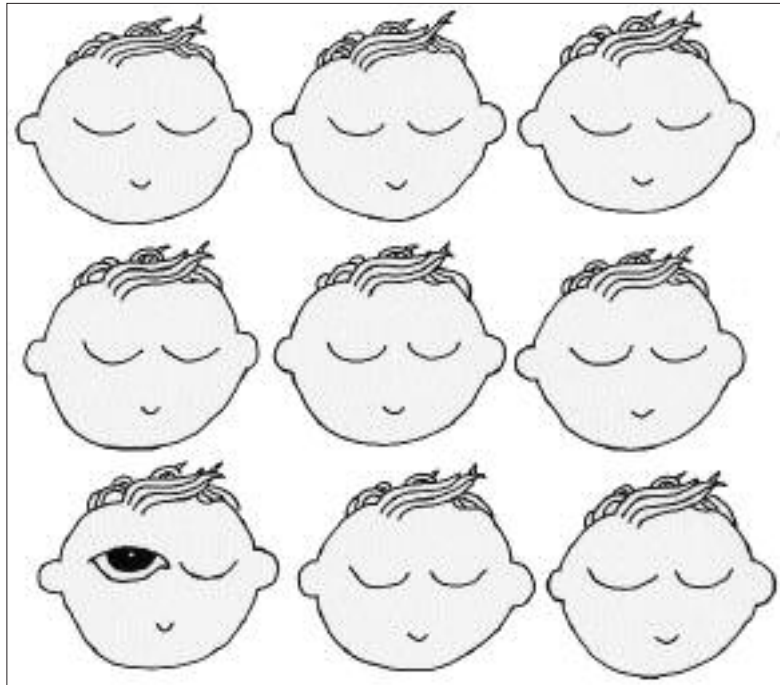
Figure 2: Transesophageal transgastric short axis view of four papillary muscles.

only at the leaflet edge and at this level the double orifice is visible. At mid and basal level the mitral valve appears normal. Our case should also be included in the incomplete bridge type, 3. Hole type: a single orifice is present at the leaflet tips, and additional smaller orifice is visible on one of the commissures oriented at a roughly right angle to main orifice (3).

No clinical signs suggest double-orifice mitral valve, electrocardiogram and chest X-ray are usually normal. Echocardiography is the method of diagnosis. Parasternal short-axis is the most useful view. Transesophageal echocardiography, so far is the best technique to define valve structure and the tensor apparatus.

## References

1. Greenfield WS. Double mitral valve. Trans Pathol Soc London 1876;27:128-9.
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**Bilin bakalım, bu çocuklardan hangisi ilerde kardiyolog olacak?!**