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Case Report

A Case of Isolated Double-orifice Mitral Valve with Normal Valve Function : An Echocardiographic Examination and Clinical Implication

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ABSTRACT

We report a case of a rare congenital anomaly, a double-orifice mitral valve, in a 19-year-old man who was admitted because of chest discomfort. Physical examination revealed no significant findings, but transthoracic echocardiography revealed 2 functional orifices of approximately equal size in the mitral valve. Color Doppler flow imaging revealed that mitral flow was normal and without regurgitation or stenosis. No other associated congenital abnormality was observed. We followed up the patient conservatively without surgical repair because both left ventricular function and mitral flow were normal. Although this patient did not require any treatment, he should be carefully followed up to find possible future pathological conditions, such as a flail leaflet causing mitral regurgitation.

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Key words : congenital heart disease, mitral valve, double-orifice mitral valve, transthoracic echocardiography

INTRODUCTION

We report a case of a rare congenital anomaly, a double-orifice mitral valve (DOMV). A DOMV is usually found to be associated with other congenital heart malformations. In this case, however, because we found no other congenital malformations an isolated DOMV was diagnosed. Transthoracic echocardiography has been useful to diagnose congenital heart malformations.

CASE REPORT

A 19-year-old man was admitted to The Jikei Daisan Hospital for the evaluation of chest discomfort. Physical examination revealed no significant findings. The discomfort of the left side of the chest occurred slightly when the patient exercised or was at rest but stopped after a few minutes and seemed unlikely to be angina pectoris or other types of pain. The patient had not previously undergone surgery.

Electrocardiography revealed normal sinus rhythm without significant ST-T changes. A chest x-ray image showed a normal cardiothoracic ratio (46%) and no abnormal cardiac shadow. The results of blood examinations were within the normal range for all values.

To screen for heart disease, we performed transthoracic echocardiography with an ultrasound system (ProSound 2, Hitachi Aloka Medical, Ltd., Tokyo, Japan). The

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long axis view and the motion mode scanning could not identify any abnormality of the position or movement of the mitral valve (Fig. 1A).

This transthoracic echocardiography showed that left ventricular (LV) diastolic and systolic dimensions were 51 mm and 31 mm, respectively. The LV ejection fraction was 68%. Spectral Doppler imaging (Fig. 1B) demonstrated normal transmitral inflow velocities (E velocity = 1.07 m/second, A velocity = 0.63 m/second, E/A = 1.70, and deceleration time = 158 milliseconds). Color Doppler imaging also demonstrated no mitral valve regurgitation through any orifice. These findings were interpreted as showing that mitral valve function was within the normal range. Scanning images of the left ventricle from the parasternal short axis view at the level of the upper mitral valve (Fig. 2) showed that the appearance of the mitral valve at the level of the valve ring and the notch formation in the center of the valve could be recognized.

Scanning at the middle level of the mitral valve (Fig. 3) showed the unexpected presence of 2 adjacent mitral orifices. The lateral orifice was located anteriorly, and the medial orifice was located posteriorly. The area of the lateral mitral orifice was 1.51 cm^2 and was similar to that the medial orifice, which was 1.58 cm^2. The image clearly demonstrated the unexpected presence of 2 adjacent mitral orifices at the level of middle part of the leaflet.

Scanning images of the lower level of mitral valve (Fig. 4) showed subvalvular structures just below the level of the

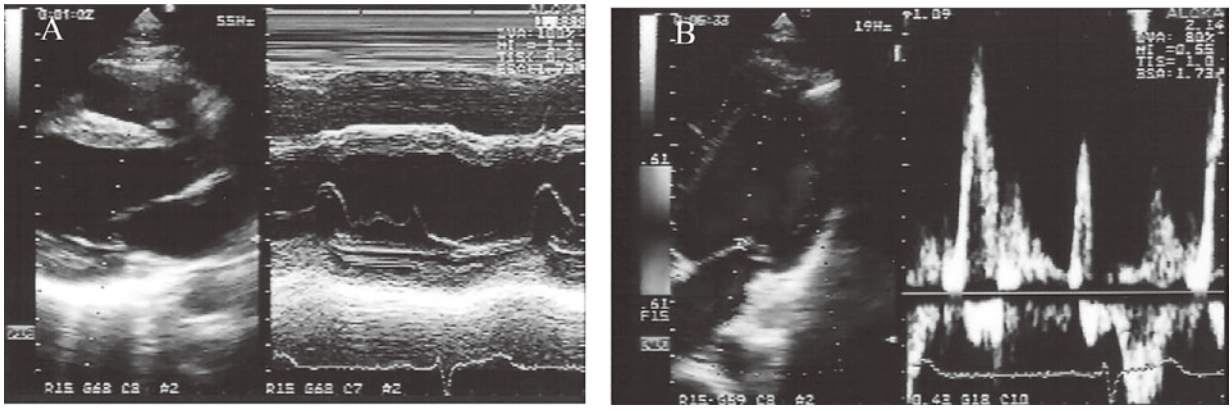
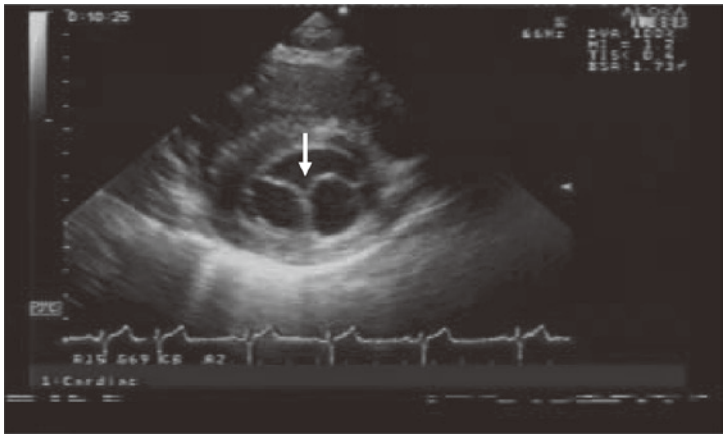
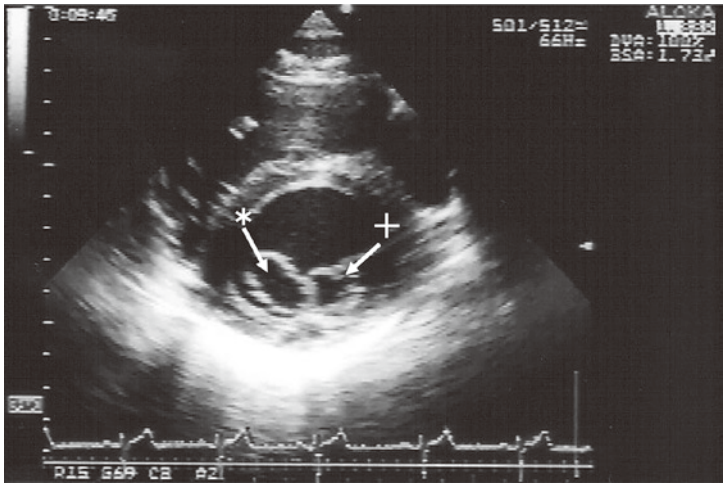


Fig. 1. A : The parasternal long axis view and M-mode echocardiography.
The appearance and motion of the mitral valve seemed to be normal.
B : Color Doppler imaging and spectral Doppler imaging of transmitral flow.
Transmitral flow showed neither a regurgitation flow pattern nor a stenotic flow pattern.



arrow : notch

Fig. 2. The upper (valve ring) level of the left ventricle from the parasternal short axis view in B mode.
A notch formation was present in the center of the mitral valve.



＋: lateral orifice
＊: medial orifice

Fig. 3. The middle (mid-leaflet) level of the left ventricle from the parasternal short axis view in B mode. Two adjacent mitral orifices were clearly observed.

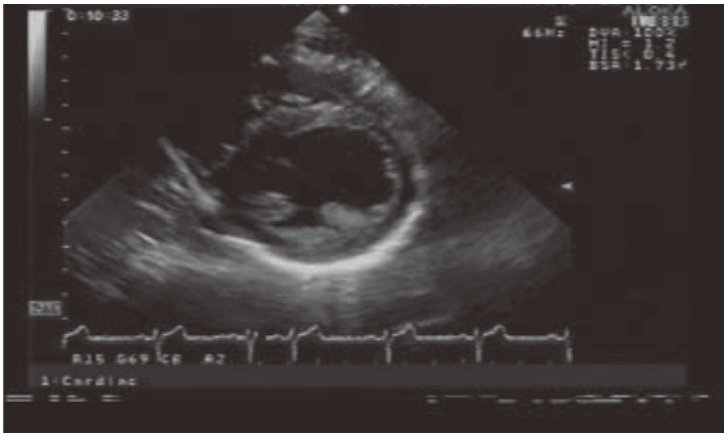


Fig. 4. The lower (leaflet edge) of the left ventricle from the parasternal short axis view in B mode. Two papillary muscles were shown.

leaflet edge. Two papillary muscles were identified, and each orifice was separately attached through the chordae to each papillary muscle.

We did not find any other cardiac malformations, such as atrioventricular septal defect. We also did not detect significant mitral stenosis or regurgitation. Therefore, we diagnosed isolated DOMV. His chest discomfort was disappeared spontaneously. So he did not need hospital treatment. We routinely follow up him every 6 month without any treatment.

Discussion

After DOMV was first described by Greenfield in 1876[1], several cases of this rare congenital anomaly have been reported. Although its exact frequency is not known, DOMV was found at autopsy in 27 (1%) of 2,733 cases of congenital heart disease[2].

Other congenital heart malformations are usually found to be association with DOMV. The malformation most commonly found is atrioventricular septal defect, and ventricular septal defect, bicuspid aortic valve, mitral regurgitation, and mitral stenosis are also found[3]. Therefore, in a case of DOMV, other congenital heart diseases should be carefully

searched for with echocardiography.

In the present case, however, we did not find any other congenital malformations. Therefore, we diagnosed this case as an isolated DOMV. The frequency of isolated DOMV is also unknown : in a previous study the DOMV was isolated in only 3 of 46 cases[4]. When this finding is combined with a DOMV rate of 1% among patients examined at autopsy[2], isolated DOMV would have occurred in, at most, 0.065% of patients examined at autopsy.

When echocardiography is performed to diagnose DOMV, it is important not to miss the partial fusion of mitral valve leaflets. The parasternal long axis view is not useful for detecting the fusion of the leaflets, and this anomaly is often missed in this view. The most useful view for demonstrating DOMV is the parasternal short axis view, which is also used to classify DOMV into 3 types, as reported by Trowitzsch et al[3]. We found the clear fusion site in the center of the mitral valve. We also detected a central fibrous subdivision with orifices of approximately equal size and 2 papillary muscles. Each orifice was separately connected to each papillary muscle. Because the 2 orifices were of almost the same size, the present case of DOMV can be classified as the complete bridge type[3].

In the present case of DOMV, neither regurgitation nor stenotic flow was detected, and no other congenital heart malformation was found. A case of isolated DOMV might have a good prognosis because of the absence of other congenital anomalies and because isolated DOMV has, in fact, been reported in an elderly patient[5]. However, several cases of congenital DOMV have been diagnosed became a flail causing severe mitral regurgitation needed to be surgically repaired[6-8]. Therefore, a patient with an isolated DOMV should be carefully followed up even if functional abnormality of the mitral valve is not present.

If we had performed routine echocardiography, we might have missed diagnosing this congenital heart malformation ; therefore, careful imaging of the mitral valve is needed to search for DOMV.

If detailed imaging of cardiac structures is obscured with transthoracic echocardiography, a more useful examination might be transesophageal echocardiography[9].

The transesophageal approach offers additional information that is useful for recognizing DOMV, evaluating its anatomic characteristics and functional status, and identifying coexisting malformations.

CONCLUSION

We have reported a case of DOMV, a rare congenital heart anomaly. Surgical repair is sometimes needed if the DOMV is associated with other congenital heart malformations. In the present case, however, no congenital heart malformation other than DOMV was found. Although this patient did not require any treatment, he should be carefully followed up to find possible future pathological conditions, such as a flail leaflet causing mitral regurgitation.

Authors have no conflict of interest.

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