Value of Three-Dimensional Echocardiography in Assessing Double-Orifice Mitral Valve in an Asymptomatic Patient

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Abstract

Double-orifice mitral valve (DOMV) is a rare congenital anomaly consisting of an accessory bridge of fibrous tissue, which divides the mitral valve (MV) into two orifices. The mitral leaflets are essentially normal in most cases, but they can be regurgitant or stenotic. It is most commonly associated with a variety of other cardiac anomalies. Isolated DOMV with normal MV function is very rare. We present here a rare case of congenital DOMV in a 25-year-old female diagnosed by real-time three-dimensional echocardiography (RT3DE). RT3DE enabled complete anatomical and functional assessment of MV apparatus. It added much valuable information over conventional 2DE that helped in establishment of the diagnosis, identification of the anatomical type, and selection of the proper management.

Keywords: Congenital heart disease, double-orifice mitral valve, real-time three-dimensional echocardiography

INTRODUCTION

Double-orifice mitral valve (DOMV) is a rare congenital anomaly that was first described by Greenfield in 1876.1 It can be seen in young and middle-aged patients, with an estimated incidence of 0.06%.2 This anomaly is consisting of an accessory bridge of fibrous tissue, which partially or completely divides the mitral valve (MV) into two orifices. The size of the orifices is balanced in only 15%.2 Anatomically and functionally, the mitral leaflets are essentially normal in most cases, but they can be regurgitant or stenotic. It is most commonly associated with a variety of other cardiac anomalies such as coarctation of the aorta, a bicuspid aortic valve, Ebstein’s anomaly, and most commonly, atrioventricular septal defects.1,3 The concomitant cardiac anomalies and malfunction of the valve usually lead to an early diagnosis in childhood. Isolated DOMV is extremely rare, but the exact incidence, prevalence, and prognostic relevance have not been established.4 DOMV with normal MV function was seen in 37% of the patients.5

We present here a rare case of congenital DOMV in a 25-year-old female diagnosed by real-time three-dimensional (3D) echocardiography.

CASE REPORT

A 25-year-old female was referred to our hospital for cardiac evaluation. She had no previous medical history. She was asymptomatic. The clinical examination was normal. Accordingly, a complete transthoracic 2D echocardiography with color Doppler examination using Philips ultrasound machine (EPIQ-7) was performed. In parasternal short-axis view, the MV orifice showed an accessory tissue between both leaflets, but it was not clearly identified as two orifices [Figure 1a]. Two papillary muscles were clearly visualized [Figure 1b]. Apical two-chamber view showed the division of MV opening into 2 by an accessory tissue. The MV was functioning well (no regurgitation or stenosis). The other valves were normal in function and morphology. To obtain a detailed description of MV apparatus, transesophageal echocardiography (TEE) was performed using the same machine and X7 matrix probe that enabled to do 2D and

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DOMV is usually detected by 2D echocardiographic examination in short-axis parasternal views. However, information obtained by 2D may be incomplete in recognition of the different types of DOMV. Some authors recommended to use magnetic resonance imaging once DOMV is nearly assured or suspected by 2D echocardiography to confirm the clinical suspicion and to exclude any concurrent cardiac and vascular abnormalities. Performing 3D echocardiography is extremely useful in identifying the morphology type and providing further anatomical and functional information, such as orifice size, number of leaflets, and its spatial relationship. In our patient, 2D transthoracic echocardiography was not helpful for assuring the diagnosis due to poor window and low image quality. Transesophageal 2D and 3D echo was used to reach the final diagnosis of DOMV, the duplicate MV type.

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DOMV can be rarely diagnosed in asymptomatic middle-aged and elderly patients, as in our case. In any case, the presence of other coexistent cardiovascular abnormalities should be excluded. The management of DOMV is related to the type and severity of MV dysfunction. Asymptomatic DOMV usually requires no active intervention; in case of severe mitral regurgitation, stenosis, or associated cardiac anomalies, surgical repair is needed. In our case, due to the absence of any abnormalities, only long-term follow-up was advised for the early detection of complications.
Conclusion

In cases of DOMV, RT3D-TEE is the most reliable method to establish the diagnosis. It also provides a comprehensive assessment of the morphology to determine the type of DOMV.

Human rights statements and informed consent

Echo procedures were performed in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1964 and later revisions. Informed consent was obtained from the patient for being included in that report.

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Conflicts of interest

There are no conflicts of interest.

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