Original article

Kardiologia Polska
2011; 69, 2: 139–143
ISSN 0022–9032

Double-orifice mitral valve — echocardiographic findings

Anna Wójcik[1], Anna Klisiewicz[1], Piotr Szymański[1], Jacek Różański[2], Piotr Hoffman[1]

[1]Department of Congenital Heart Diseases, Institute of Cardiology, Warsaw, Poland
[2]2nd Department of Cardiac Surgery, Institute of Cardiology, Warsaw, Poland

Abstract

Background: Double-orifice mitral valve (DOMV) is a rare congenital malformation characterised by the presence of two orifices in the left atrio-ventricular valve area, each having an independent chordal attachment to the papillary muscle.

Aim: To establish the echocardiographic features and incidence of DOMV in patients undergoing an echo study in a tertiary referral centre.

Methods: We carried out a retrospective review of 215,193 echocardiographic studies in 79,919 patients performed between 1993 and 2006. Transthoracic echocardiographic examinations (TTE) of nine patients with DOMV (six female, mean age 37.9 years, range 8–59) were analysed.

Results: In six patients, the complete bridge type of DOMV was recognised, two patients had a duplicate mitral valve, and one had the hole type. In all cases, mild to moderate mitral regurgitation was present. Moderate to severe mitral stenosis was diagnosed in five cases and was associated with the complete bridge type. In four patients, ostium primum atrial septal defect was present. The TTE was sufficient to define the type, anatomy and associated lesions in seven cases. In two patients, TEE was essential to establish the diagnosis.

Conclusions: Transthoracic echocardiographic examination is a reliable, and in most cases sufficient, means of diagnosing DOMV and determining its type. The DOMV as a cause of symptomatic mitral valve disease is seen in middle-aged/elderly patients. The incidence of double-orifice mitral valve in the adult tertiary referral echocardiographic laboratory is estimated at 0.01%.

Key words: DOMV, mitral valve, ASD primum, heart septal defects

Kardiol Pol 2011; 69, 2: 139–143

INTRODUCTION

Double-orifice mitral valve (DOMV) was described by Greenfield in 1876. To date, approximately 200 cases have been reported [1–3]. This very rare malformation is characterised by the presence of two orifices in the left atrio-ventricular (AV) valve area, each having an independent chordal attachment to the papillary muscle. It is thought to result from abnormal fusion of the endocardial cushions and abnormal development of the mitral valve from the primary fold and primitive left ventricle during a delamination process [4].

A DOMV can occur as an isolated anomaly, but in most cases it is associated with other congenital malformations. The clinical presentation of DOMV varies and depends mainly on the associated lesion [5] and degree of mitral stenosis (MS) and/or mitral regurgitation (MR) [6].

Various classifications for DOMV have been proposed, based on the size and location of the two orifices [4]. Trowitz et al. [5] described three different types of this anomaly using 2D echocardiography: (1) complete bridge type (about 15% of DOMVs), in which both orifices are visible from the

Address for correspondence:
Anna Wójcik, MD, Department of Congenital Heart Diseases, Institute of Cardiology, ul. Alpejska 42, 04–628 Warszawa, Poland,
tel: +48 22 343 44 57, fax: +48 22 343 45 21, e-mail: a.wojcik@ikard.pl
Received: 30.06.2010 **Accepted:** 24.11.2010

leaflet edge, all the way through the valve ring. Both openings are circular (equal or unequal in size), papillary muscles usually are normal, with chordae surrounding each orifice inserting into one papillary muscle; (2) incomplete bridge type, in which the connection is seen only at the leaflet edge, resulting in a double circle only at the leaflet level, with a normal appearance in the more basal views, and (3) hole type — the commonest variety, characterised by a small accessory orifice situated at either the anterolateral or posteromedial commissure, visible only at the mid-leaflet level and disappearing on scanning toward the apex or base. When the accessory orifice is located at the posteromedial commissure, a common AV canal usually is present [3, 6, 7]. Some authors described a fourth type of DOMV: duplicate mitral valve with two annuli and valves, each with its own set of leaflets, commissures, chordae, and papillary muscles [6].

The purpose of our study was to analyse the function and anatomy of DOMV and its association with other congenital lesions in a group of nine consecutive patients with DOMV detected by echocardiography in our laboratory.

METHODS

We carried out a retrospective review of nine patients with DOMV diagnosed between 1993 and 2006 in our echocardiographic laboratory (six females, mean age 37.9 years, range 8–59). Patients had been referred for echocardiographic evaluation with an initial diagnosis of mitral valve disease (five patients) or congenital heart disease with left-to-right shunt (four patients). Transthoracic studies (TTE) were performed in all patients. In addition, two patients underwent transesophageal examination (TEE) due to equivocal TTE results. Two-dimensional images of the parasternal long- and short-axis views, apical four- and two-chamber views were inspected for the location and the size of the orifices. The DOMV was classified based on valve morphology visualised in the different short-axis planes (from the leaflet edges all the way through the valve ring, at the leaflet edge level, at the mid-leaflet level). The echocardiographic study was completed by comprehensive Doppler imaging (evaluation of the systolic pressure gradient across the mitral valve, the mitral valve area and the degree of MR). Echocardiographic data were compared and contrasted with that generated by direct surgical inspection.

RESULTS

In six patients, the complete bridge type DOMV was detected (Figs. 1, 2). Two other patients had the duplicate mitral valve type (Fig. 3), and one had the hole type. In all cases, mild to moderate MR was present. Moderate to severe MS was diagnosed in five cases and was associated with the complete bridge type (Fig. 4). In four patients, the ostium primum atrial septal defect (ASD) was present (one of the complete bridge type, two of the duplicate MV type, one of the hole

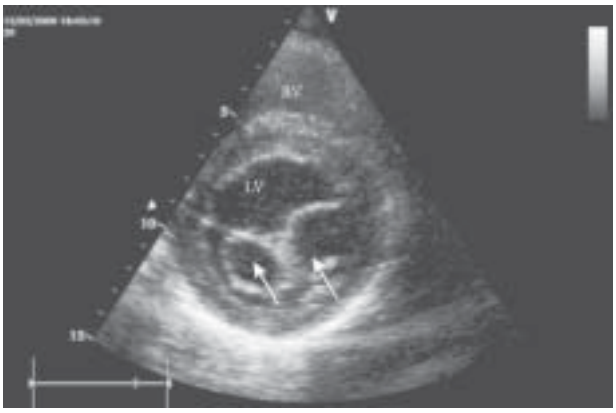

Figure 1. Short-axis transthoracic view. Complete bridge type of double-orifice mitral valve (arrows); RV — right ventricle; LV — left ventricle

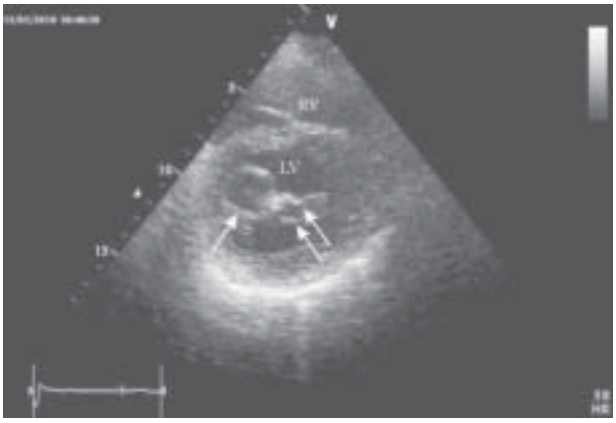

Figure 2. Short-axis transthoracic view. Complete bridge type of double-orifice mitral valve. The orifices (arrows) unequal in size; RV — right ventricle; LV — left ventricle

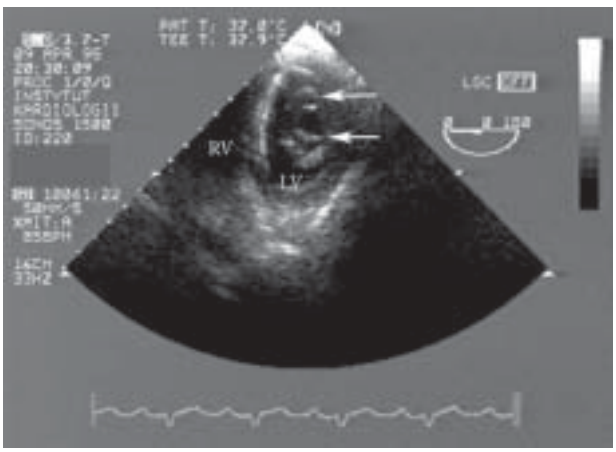

Figure 3. Transesophageal view showing duplicate double-orifice mitral valve (arrows); RV — right ventricle; LV — left ventricle

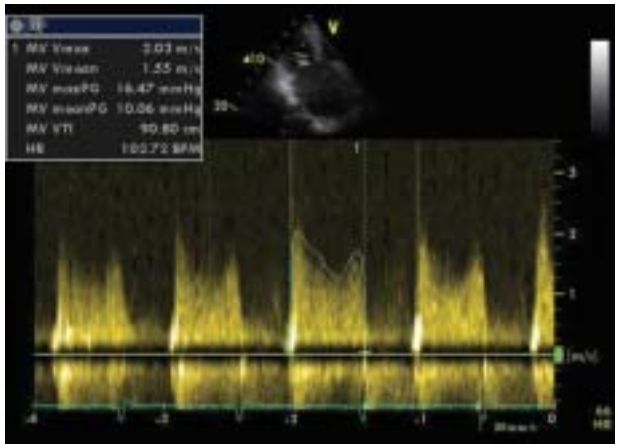

Figure 4. Apical transthoracic view. Increased transmitral systolic pressure gradient

type). In three cases, severe (and in another two mild) tricuspid regurgitation was observed. Three patients had mild aortic regurgitation. The TTE was sufficient to define the type, anatomy and associated lesions in seven cases. Because in the other two patients TTE was equivocal, TEE was needed to establish the diagnosis. Five patients were operated on. In the group of four patients with ASD primum, the atrial defect was closed with a patch; in two cases the smaller orifice was closed. In three cases, the cleft of the anterior mitral leaflet was sutured. One patient with DOMV as an isolated anomaly had mitral valve replacement. Post-operative recovery was uneventful. During the long-term follow-up of patients after the operation of ASD primum and mitral valvuloplasty, moderate MR was present in two patients, mild in one and severe in the fourth.

The demographic data, anatomical and functional characteristics of the DOMV in the studied group are summarised in Table 1.

DISCUSSION

The clinical presentation of DOMV is variable and mostly depends on the associated lesions [3] and the functional status of both orifices. The function of both components of the DOMV can be normal but is mildly impaired in about half of patients. Mild to severe regurgitation or significant stenosis of either or both orifices has been reported [2]. The DOMV can be an isolated anomaly, but in most cases it is associated with other congenital malformations such as AV canal defect, ventricular septal defect, coarctation of the aorta, interrupted aortic arch, subaortic stenosis, patent arterial duct and primum type ASD. Occasionally, DOMV coexists with secundum type ASD, tetralogy of Fallot, hypoplastic left heart syndrome, Ebstein's anomaly or bicuspid aortic valve [4].

Mitral regurgitation is the commonest functional abnormality, and MS, either alone or with insufficiency, is rare [3, 4]. In our group, all the patients had mild to moderate MR, and five out of the nine also had significant MS, associated with the complete bridge type of DOMV, which was the commonest variety in our group. The hole type of DOMV is thought to be the commonest type (more than 80% of patients) and is often associated with a common AV canal [6]. In our group, the hole type was diagnosed in only one patient and was accompanied by the ASD primum type. In two other patients with ASD primum, the duplicate form of DOMV was present, and in one, complete bridge type was present.

In one of the largest groups (46 children) with DOMV described by Zalzstein et al. [3], MR was the most frequent finding (43%) whereas MS was present in 13% of cases, and combined MS and insufficiency — in 6.5% of patients. The most commonly associated anomaly was AV septal defect (39%), which is similar to previous findings. Obstructive left sided lesions were present in 41% of cases (coarctation of the aorta, subaortic stenosis). Other anomalies were rare (transposition of the great arteries, ventricular septal defect, truncus arteriosus, tricuspid atresia, Ebstein's anomaly, double

Table 1. Demographic data, anatomical and functional characteristics of double-orifice mitral valve in the studied group

Patient	Age	Gender	Type of DOMV	MR	MS pressure gradient/MVA	Associated lesions
1	17	M	Complete bridge	Mild	19/9.5 mm Hg/1.7 cm^2	Severe TR, mild AR
2	15	M	Complete bridge	Moderate	14.3/8.2 mm Hg/1.1 cm^2	–
3	56	F	Complete bridge	Mild	16/4.1 mm Hg/1.5 cm^2	Mild TR
4	8	F	Hole	Mild	–	ASD I
5	59	F	Duplicate	Moderate	–	ASD I, severe TR
6	15	F	Complete bridge	Moderate	–	ASD I
7	59	F	Complete bridge	Moderate	14.4/6 mm Hg/0.9 cm^2	Moderate TR, mild AR
8	54	M	Duplicate	Mild	–	ASD I, mild TR
9	58	F	Complete bridge	Mild	12.8/4.6 mm Hg/1.5 cm^2	Mild AR

DOMV — double-orifice mitral valve; MR — mitral regurgitation; MVA — mitral valve area; M — male; F — female; MS — mitral stenosis; TR — tricuspid regurgitation; AR — aortic regurgitation; ASD I — primum type atrial septal defect

outlet right ventricle with ventricular septal defect and pulmonary stenosis) [3].

There are no specific clinical signs suggestive of DOMV [3]. None of the patients was referred to our echo lab with an initial diagnosis, or even a suspicion, of DOMV. Five of them had had mitral valve disease diagnosed in the past, and four were examined because of primum type ASD. The DOMV is a very rare anomaly, but one should be aware of its presence when examining patients with mitral valve disease and ASD primum type.

Of particular interest are five patients with DOMV diagnosed at the age > 50 years in our series, because although since 1952 about 100 papers reporting double-orifice mitral valve have been published, only one case of a patient aged over 50 has been described [8–11].

The true incidence of DOMV is yet to be established. In our database, there were 215,193 echocardiographic studies of 79,919 patients performed between 1993 and 2006 which produced an incidence of double-orifice mitral valve in our centre of approximately 0.01%. This is a much lower rate than that recorded at the Children's Hospital, Denver, USA, where 18 patients with an intact AV septum were identified from 40,179 echocardiographic studies performed between 1997 and 2002 [12]. This however can be easily explained by the different clinical profile of a paediatric and an adult echocardiographic laboratory.

CONCLUSIONS

1. Transthoracic echocardiographic examination, especially in short-axis parasternal views, is a reliable method and in most cases sufficient to confirm a diagnosis of double-orifice mitral valve and to determine its type.
2. Double-orifice mitral valve as a cause of symptomatic mitral valve disease is also seen in middle-aged/elderly patients.
3. The incidence of the diagnosis of double-orifice mitral valve in the adult tertiary referral echocardiographic laboratory is 0.01%.

Conflict of interest: *none declared*

References

1. Purnode P, Rombaut E, Gerard M et al. Double orifice mitral valve with flail leaflet: a transesophageal echocardiographic examination. Eur J Echocardiogr, 2000; 1: 144–146.
2. Hoffman P, Stumper O, Groundstroem K et al. The transesophageal echocardiographic features of double-orifice left atrioventricular valve. J Am Soc Echocardiogr, 1993; 6: 94–100.
3. Zalzstein E, Hamilton R, Zucker N et al. Presentation, natural history, and outcome in children and adolescents with double orifice mitral valve. Am J Cardiol, 2004; 93: 1067–1069.
4. Erdemli O, Ayik I, Karadeniz U et al. A double-orifice atrioventricular valve case: intraoperative tranesophageal echocardiography in diagnosis and treatment. Anesth Analg, 2003; 97: 650–653.
5. Trowitz E, Bano-Rodrigo A, Burger BM et al. Two-dimensional echocardiographic findings in double orifice mitral valve. J Am Coll Cardiol, 1985; 6: 383–387.
6. Wójcik A, Klisiewicz A, Lusawa T et al. Double-orifice mitral valve: case report. Kardiol Pol, 2005; 63: 663–665.
7. Jose VJ, Chandy ST, John B. Double orifice mitral valve. Indian Heart J, 2003; 55: 279–280.
8. Kron J, Standerfer RJ, Starr A. Severe mitral regurgitation in a woman with a double orifice mitral valve. Br Heart J, 1986; 55: 109–111.
9. Heyse AM, Vanhercke D, Nimmegeers J et al. Mitral insufficiency with congenital double-orifice mitral valve in an elderly patient. Eur J Echocardiogr, 2003; 4: 334–335.
10. Kim SJ, Shin ES, Lee SG. Congenital double-orifice mitral valve with mitral regurgitation due to flail leaflet in an elderly patient. Korean J Intern Med, 2005; 20: 251–254.
11. Sasaoka T, Ohguri H, Makita Y et al. Double-orifice mitral valve in an elderly patient with tetralogy of Fallot. Jpn Heart J, 1996; 37: 503–507.
12. Das BB, Pauliks LB, Knudson OA et al. Double-orifice mitral valve with intact atrioventricular septum: an echocardiographic study with anatomic and functional considerations. J Am Soc Echocardiogr, 2005; 18: 231–236.

Dwuujściowa zastawka mitralna — charakterystyka echokardiograficzna

Anna Wójcik[1], Anna Klisiewicz[1], Piotr Szymański[1], Jacek Różański[2], Piotr Hoffman[1]

[1]Klinika Wad Wrodzonych Serca, Instytut Kardiologii, Warszawa
[2]II Klinika Kardiochirurgii, Instytut Kardiologii, Warszawa

Streszczenie

Wstęp: Dwuujściowa zastawka mitralna (DOMV) jest wrodzoną, rzadką anomalią charakteryzującą się obecnością 2 oddzielnych otworów w obrębie lewego ujścia przedsionkowo-komorowego.

Cel: Celem badania było określenie typu morfologicznego i częstości występowania DOMV wśród pacjentów diagnozowanych w pracowni echokardiograficznej, w której pracują autorzy niniejszej pracy.

Metody: Autorzy dokonali retrospektywnego przeglądu 215 193 zapisów echokardiograficznych z badań przeprowadzonych u 79 919 pacjentów w latach 1993–2006.

Wyniki: Obecność DOMV stwierdzono u 9 chorych (śr. wiek 37,9 roku, zakres 8–59 lat, w tym 6 kobiet). U wszystkich rejestrowano różnego stopnia niedomykalność mitralną, u 5 zwężenie ujścia, a u 4 pacjentów wadzie towarzyszył ubytek przegrody międzyprzedsionkowej typu pierwszego. Echokardiograficzne badanie przezklatkowe pozwoliło postawić pełną diagnozę u 7 pacjentów, zaś u pozostałych 2 osób było konieczne badanie przezprzełykowe.

Wnioski: Echokardiograficzne badanie przezklatkowe jest wiarygodnym badaniem diagnostycznym, które w większości przypadków wystarcza do rozpoznania DOMV i określenia typu tej anomalii. U osób w średnim lub podeszłym wieku DOMV może być przyczyną objawowej choroby zastawki mitralnej. Częstość występowania DOMV w ośrodku autorów wyniosła 0,01%.

Słowa kluczowe: dwuujściowa zastawka mitralna, ubytek przegrody międzyprzedsionkowej typu pierwszego

Kardiol Pol 2011; 69, 2: 139–143

Adres do korespondencji:
lek. Anna Wójcik, Klinika Wad Wrodzonych Serca, Instytut Kardiologii, ul. Alpejska 42, 04–628 Warszawa, tel: +48 22 343 44 57, faks: + 48 22 343 45 21, e-mail: a.wojcik@ikard.pl
Praca wpłynęła: 30.09.2010 r. **Zaakceptowana do druku:** 17.11.2010 r.