

Diagnosis and Management of the Double Orifice Mitral Valve: Three Case Reports

Omer Faruk Dogan, MD,¹ Metin Demircin, MD,¹ Tevfik Karagoz, MD²

¹Department of Cardiovascular Surgery; ²Department of Pediatri, Division of Pediatric Cardiology, Hacettepe University Medical Faculty, Ankara, Turkey



Dr. Dogan

ABSTRACT

Double orifice mitral valve (DOMV) is an uncommon congenital heart defect. The isolated occurrence of this anomaly is very rare and, more often, is associated with another congenital malformation, dominated by atrioventricular canal defects (AVCD). Mitral insufficiency and/or stenosis may complicate this malformation. Treatment may be summarized as abstention, surgical valve repair, or valve replacement.

In here, we report three cases with DOMV. The first patient was an 18-month-old boy who was operated for severe mitral valve stenosis and mild insufficiency (in 1980s), the second was a 47-year-old female, who was operated recently for mitral valve replacement (MVR) under cardiopulmonary bypass due to severe mitral valve insufficiency. This patient had been operated due to the secundum-type atrial septal defect in our institution previously. The last one, a 3-year-old boy, with DOMV and three papillary muscles, was on clinical follow-up because he had no symptoms.

INTRODUCTION

The double orifice mitral valve (DOMV) was first described by Greenfield in 1876 [Bano-Rodrigo 1988] and it is uncommon but a surgically and clinically important condition. It has been defined as associated most often with AVCD, although it may occur as an isolated malformation or in association with other cardiac anomalies.

DOMV or duplication of the mitral valve usually presents a congenital mitral stenosis or may show a normal valve function [Saylam 1976]. This malformation of the valve sometimes presents with no hemodynamic disturbances, but usually produces mitral valve stenosis and very rarely mitral

insufficiency [Yurdakul 1995]. In half of the reported cases, the duplication of the mitral valve occurs as an isolated lesion [Rosenberg 1986].

We would like to present three cases of DOMV. Two of our patients were operated under cardiopulmonary bypass because of severe mitral valve stenosis and insufficiency and the other was on medical follow-up. The mitral valve is supported by the chorda tendinea originating from two papillary muscles in two patients, and three papillary muscles and approximately equal size of mitral orifices with normal valvular function was recorded by means of the transthoracic echocardiography in the other.

CASE I

An 18-month-old boy was referred to the division of pediatric cardiology due to recurrent upper airway infection. He also had cyanosis of the fingertips and the mouth. Engorged cervical veins were observed on inspection. Physical examination revealed normal blood pressure and sinus tachycardia. Precordial thrill, apical holosystolic murmur were noted and the liver was palpable about 3 cm below the costal margin. Chest x-ray revealed global cardiac enlargement (Figure 1), and increased pulmonary vascularity. Electrocardiography showed left atrial dilatation and left ventricular hypertrophy. Severe mitral valve regurgitation was recorded during cardiac catheterization when the contrast medium was injected into the left ventricle. The patient was given digitalis and diuretics in the preoperative period, and mitral valve replacement was planned.

The operation was performed by use of the cardiopulmonary bypass (CPB) and under hypothermia of 25°C. Left atrium was opened and mitral valve was visualized. Mitral valve showed two orifices which were unequally separated by a fibrotic band, one of 1 cm and the other 3.5 cm in diameter (Figures 2 and 3). There were two underlying papillary muscles. Mitral valve was incompetent and deformed. The mitral valve was excised and replaced by the artificial Lillehei-Kaster mitral valve prosthesis (No: 20) using continuous suture. He did well during the postoperative period and was kept on antiaggregant and anticoagulant therapy for the prevention of thromboembolic events. He was also on a dose of digitalis but seldom diuretics. Control chest x-

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Address correspondence and reprint requests to: Omer Faruk Dogan, MD, Birlik Mabellesi, 59.Sokak, 9/1, 06670, Ankara, Turkey (e-mail: ofdogan@hacettepe.edu.tr).

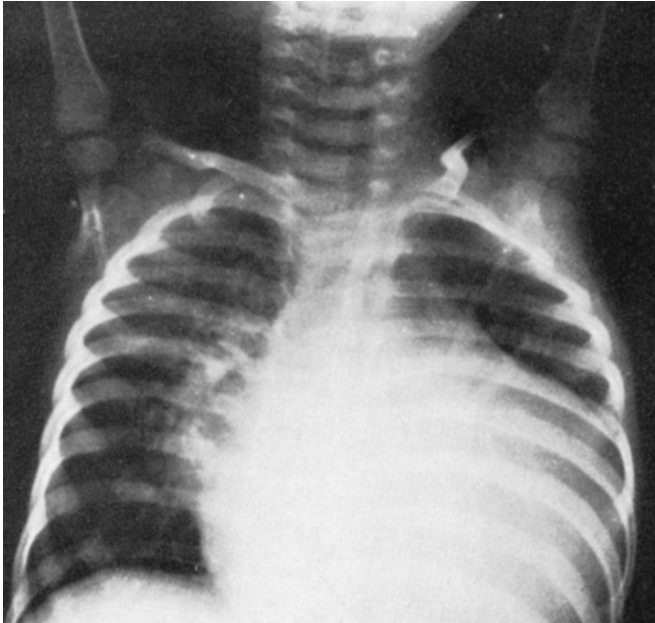


Figure 1. Preoperative chest x-ray shows global enlargement of the heart.

ray during the postoperative phase showed a cardiac silhouette diminished in size compared to the preoperative period (Figure 3). He discharged from the hospital when his condition was good.

CASE 2

A 46-year-old female patient came to the hospital for the first time when she was 4 years old, complaining of effort dyspnea and exercise intolerance. Echocardiography revealed a diagnosis of secundum-type ASD. ASD was closed

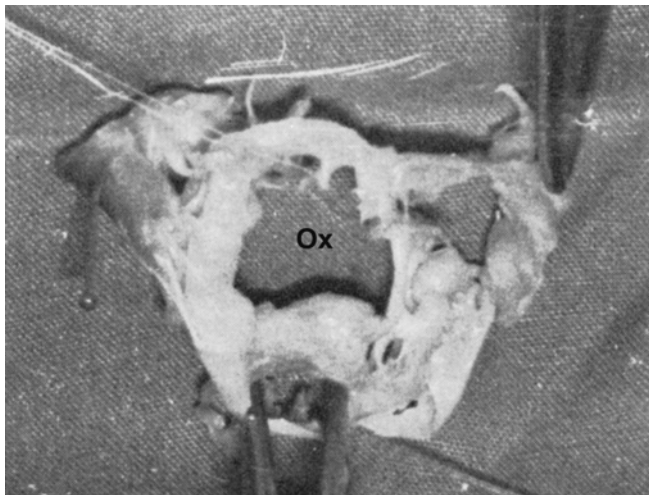


Figure 2. The unequal size of the two orifices of the mitral valve which is harvested for pathologic specimen. Ox indicates the large anterior orifice.

primarily under hypothermic CPB, and the patient was discharged with excellent postoperative progress. After 3 years, she was admitted to Hacettepe University Cardiovascular Surgery Clinic due to dyspnea on effort and palpitations. Thus, the patient was diagnosed with congenital heart disease, probably a mitral stenosis. Accordingly, she was given medical treatment. Her treatment over the following years was limited to the control of dyspnea. Then, an echocardiographic examination in 1972 revealed that the mitral valve might have double orifices. But, mitral stenosis was mild and symptoms disappeared with medical treatment. She was followed up for almost 32 years because of moderate mitral stenosis. In 1994, the patient was admitted to hospital with refractory congestive heart failure. The diagnosis of DOMV was established during this admission by means of the transthoracic ECHO. Moderate mitral stenosis and an insufficiency of 1-2 degree were detected in both mitral valves. In the same year, she gave normal birth. Following her discharge from the hospital, the patient's cardiac status was regularly checked by echocardiography. In 2001, mitral valve was replaced due to severe mitral stenosis and a second-level mitral insufficiency was diagnosed by color-Doppler echocardiogram and left cardiac catheterization. Peak diastolic gradient was 24 mm Hg (mean was 12 mm Hg). Cardiac catheterization noted a peak pulmonary capillary wedge pressure of 12 mm Hg, pulmonary artery systolic pressure was 35-40 mm Hg.

The patient underwent surgical correction. The two orifices of the mitral valve which was supported by chordae tendineae originating from the papillary muscles and atrioventricular wall were unequal in size, the major orifice was

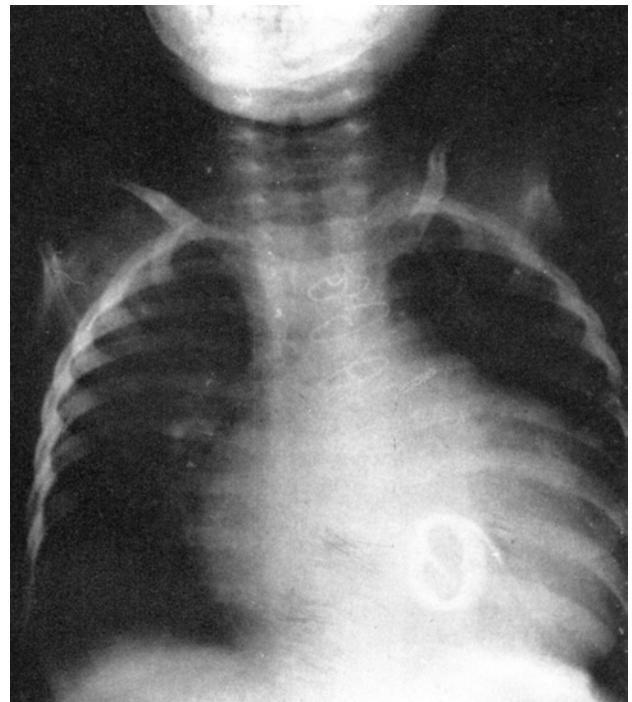


Figure 3. Thoracic chest x-ray showing the artificial heart valve and decrease in the cardiac silhouette postoperatively.

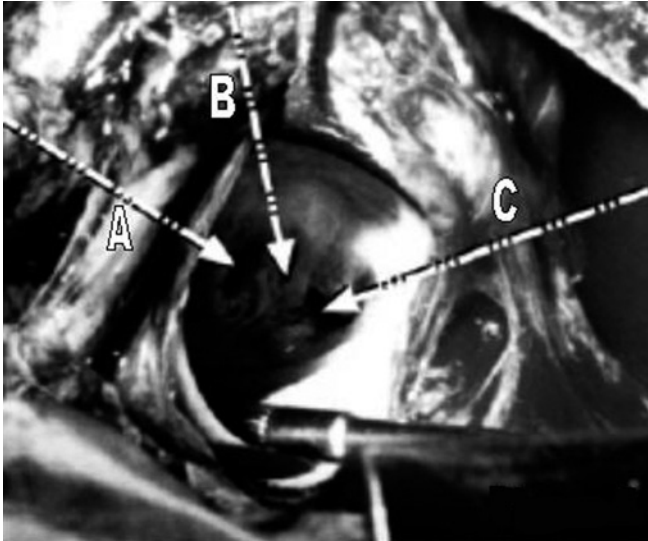


Figure 4. Unequal size of the two orifices is shown in this illustration. Picture of the double orifice mitral valve is seen through the left atriotomy intraoperatively. Arrows A, B, and C indicate the small orifice, bridging tissue, and the larger orifice, respectively.

located anteriorly and the other one posteriorly (accessory orifice) (Figure 4). The DOMV completely separated two unequal size orifices by a bridge. The valve was calculated as .4 cm in the anterolateral orifice and 1.34 cm in diameter in the posterolateral orifice. The valvular cusps of the main or the accessory mitral valve had proper chordae tendinae and papillary muscles. Several semi-regular (partially normal) chordae tendinae which were originating from anterolateral and posteromedial leaflets were attached to anterolateral-middle and posteromedial-middle papillary muscles respectively. In addition, mitral valve was highly fibrotic. It also showed examples of severe stenosis and mild insufficiency. Thus, the excised valve was replaced with no. 31 St. Jude mechanical valvular prosthesis with use of cardiopulmonary bypass with moderate hypothermia (28°C). She weaned from CPB successfully. Postoperative course was uneventful and the patient discharged from the hospital 7th day postoperatively.

CASE 3

A 3-year-old male patient was referred to the division of the pediatric cardiology because of a recurrent high body temperature. Abnormal physical findings were limited to the heart. There were no murmur or electrocardiographic abnormality. He complained of palpitation sometimes only. With a transthoracic short axis view of the two-dimensional echocardiogram, the two orifices were evident at the level of the mitral valve (Figure 5). While sweeping the probe from the apex toward the base of the heart, both anterolateral and posteromedial orifices were clearly visible from the leaflet edge all the way through the valve ring. Each orifice was nearly circular and approximately equal in size (Figure 6). In addition, three papillary muscles were revealed (Anterior, posterior and the medial papillary muscles) (Figure 7). The func-



Figure 5. Subcostal short-axis view of a double orifice mitral valve. One orifice is superior and the other inferior. The orifices are almost equal in size.

tion of this DOMV was essentially normal, and no findings of mitral stenosis were obtained. There was also no evidence of other cardiac defect such as AVCD, patent ductus arteriosus or ASD. He was discharged from the hospital for clinical follow-up. He was in a normal cardiac and exercise status after 6 months diagnosis.

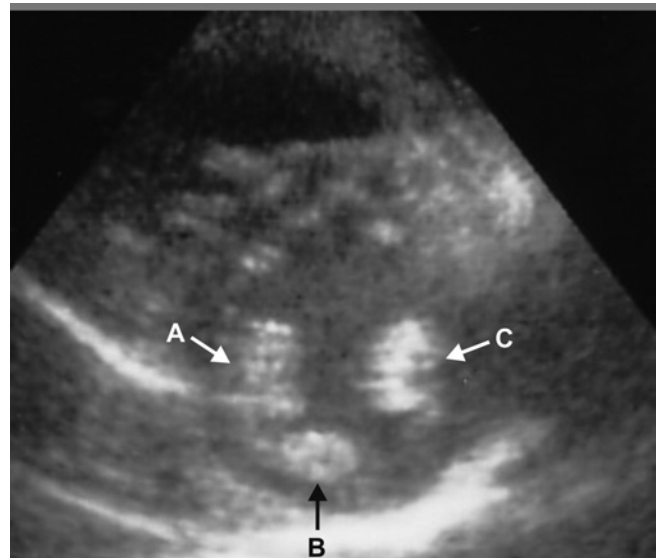


Figure 6. Parasternal short-axis view of left ventricle demonstrating three papillary muscles. Arrow A: the anterior papillary muscle, Arrow B: the middle papillary muscle, and Arrow C: the posterior papillary muscle.

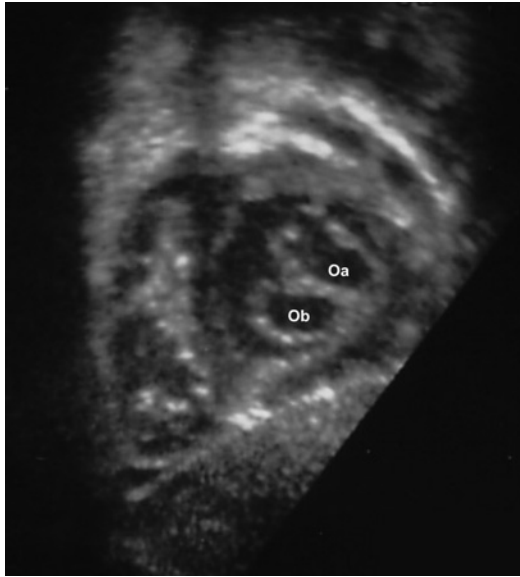


Figure 7. A modified apical four-chamber view of the heart showing a fibrous bridge between two leaflets dividing the mitral valve into two equal size orifices. Oa and Ob are clearly demonstrating the approximately equal size of the orifices of the mitral valve by means of the transthoracic echocardiography.

DISCUSSION

Anatomic as well as hemodynamic observations have shown that DOMVs function as competent units. Echocardiographic technique has allowed a noninvasive and more frequent detection of this entity. The DOMV and additional lesions of each subdivision of the mitral valve were clearly shown by two-dimensional and color-Doppler echocardiography [Segni 1986]. Of all the present cases with DOMV were diagnosed with transthoracic color-doppler echocardiography only at the hospital admission. Because techniques of echocardiogram and experiences were not enough to be shown this anomaly with anatomical details before 1980. Most cases of DOMV were described intraoperatively or postmortem incidentally in the literatures.

Double mitral valve orifice may occasionally be of functional significance causing mitral stenosis or incompetence and usually associated with other cardiac anomalies [Demircin 2004, Domenico 1993, Honnekeri 1993], mainly endocardial cushion defect [Ancalmo 1977]. In 25% of the reported cases, partial atrioventricular septal defect is associated with this malformation, suggesting that the developmental defect involves endocardial cushion tissue [Yurdakul 1995]. However, in the second case, there was only a fossa ovalis type atrial septal defect as associated anomaly, which was closed in the first operation, and the other had no cardiac defect or malformation except DOMV.

In this anomaly the valve leaflet of two orifices found in the left atrioventricular orifice is supported by chordae tendinae originating from two papillary muscles; however, only one case with DOMV and three papillary muscles have been reported by Hashimoto in 1993 [Hashimoto 1993]. To our

knowledge, one of our three cases, who is presented, is the second case with DOMV and without other cardiac anomalies and symptoms in English literature. Amano and Suzuki performed division of the fibrous bar and value reconstructive by cleft suture and leaflet expansion [Amano 1986].

The absence of associated cardiac anomalies is more frequent in patients with DOMV with equal orifices. But, most patients who have been presented are infrequent in the literature. DOMV unassociated with other cardiac defects, however, are very rare and usually live a healthy life.

According to Bano-Rodrigo et al. (1988), the appearance of equal-sized orifices is limited to 15% of patients with the DOMV anomaly. Other cases have shown unequal orifices in the two valves. Indeed, DOMV was divided into two equal orifices by a bridged tissue in our last patient, and he had no cardiac symptoms.

In 48% of patients with DOMV, the mitral valves function normally, while mitral stenosis is documented in 26% regurgitation is also seen in 26%. In the first case, thus, having such as this deformity, the patient giving a normal birth was unbelievable. However, the second patient had no mitral insufficiency or stenosis.

The framework that composes the DOMV can be divided into three forms: a complete bridge between the two valves, an incomplete bridge, and a hole formed by the congenitally abnormal valves. A complete bridge is the structure that is least commonly seen like in this case [Sasaoka 1966].

It is now accepted that the true developmental basis of duplication of the mitral valve is unknown [Rosenberg 1986]. In the only study having a logical hypothesis for the time being, Lewis, after studying the Harvard collection of pig embryos, proposed that the double mitral valve was the result of an early arrest of development and that the accessory orifice represents a retention of the left portion of the common atrioventricular canal with subsequent reduction of the mitral ostium and alignment with it [Demircin 2004].

The non-cleft orifice of a DOMV usually is competent and rarely requires closure. The cleft, because it constitutes a type of parachute (single papillary muscle) valve, should be closed partially so as to relieve valve incompetence without causing undue stenosis. Repair of atrioventricular canal associated with DOMV can be achieved with a low operative mortality and excellent late results [Lee 1999]. Reed et al (1970) were first described as a successful surgical treatment for DOMV in their case with primum-type ASD and cleft mitral. It may cause peculiar surgical problems when plastic repair of the mitral valve is needed [Segni 1986]. Treatment may be summarized as abstention, surgical repair, or valve replacement. In the present case, MVR (metallic) was preferred because of fibrocalcific valves, mitral valve stenosis with insufficiency [Lee 1985].

If the patients had a DOMV with unequal size orifices, generally they had together with other cardiac malformations and they need a surgical corrective operation. Indeed, we performed surgical operation under CPB in our two patients with DOMV with unequal size orifices.

Tomita et al (1997) have reported a patient with DOMV and torn chordae, who was successfully treated with artificial cordae replacement. Cases with equal orifices are less frequent [Bano-Rodrigo 1988, Reed 1970, Rosenberg 1968]. Bano-

Rodrigo et al (1988) reported in their postmortem study that only four cases had equal orifices due to presence of a central fibrous subdivision, which may restrict the left ventricular inflow and may contribute to the hypoplasia of the left heart. Our patient had no hypoplastic left heart anomaly.

In conclusion, DOMV is an unusual anomaly not generally associated with hemodynamic deficits. It is frequently associated with other cardiovascular anomalies such as ASD and AVCD. Echocardiographic (transthoracic or transesophageal) examination is an easy, safe, and non-invasive method, and it gives detailed information about the valve anatomy, and associated with other cardiac defects in the same time. In our opinion, different corrective methods have been described, although the patient with DOMV is an asymptomatic surgical procedure is no need and clinical follow-up is enough. Also, MVR may be performed reliably under hypothermic CPB in patients with DOMV and together with severe valvular insufficiency and/or stenosis, especially when mitral valve repair could not be performed or has an unsuccessful result.

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