CASE REPORT

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Double orifice mitral valve – A case report

Dvostruki orificijum mitralne valvule

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Abstract

Introduction. Double orifice mitrol valve (DOMV) is a very rare congenital heart defect. **Case report.** We reported 20-year-old male referred to our center due to evaluation of his cardiologic status. He was operated on shortly after birth for a tracheoesophageal fistula. Accidentally, echocardiography examination at the age of 4 years revealed double orifice mitral valve (DOMV) without the presence of mitral regurgitation, as well as mitral stenosis, with normal dimensions of all cardiac chambers. The patient was asymptomatic, even more he was a kick boxer. His physical finding was normal. Electrocardiography showed regular sinus rhythm, incomplete right bundle branch block. Transthoracic echocardiography (TTE) examination revealed the normal size of the left atrial, mitral leaflets were slightly more redundant. The left and right heart chambers, aorta, tricuspid valve and pulmonary artery valve were normal. During TTE examination on a short axis view two asymmetric mitral orifices were seen as a double mitral orifice through which we registered normal flow, without regurgitation and mitral stenosis. Transesophageal echocardiography (TEE) examination from the transgastric view at the level of mitral valve, showed 2 single asymmetric mitral orifices separated by fibrous tissue, mitral leaflet with a separate insertion of hordes for each orifice. **Conclusion.** The presented patient with DOMV is the only one recognized in our country. The case is interesting because during 16-year a follow-up period there were no functional changes despite the fact that he performed very demanded sport activities. This is very important because there is no information in the literature about that.

Key words:
heart defects, congenital; diagnosis; echocardiography, transesophageal.

Apstrakt

Uvod. Dvostruki orificijum mitralne valvule (DOMV) je vrlo retka kongenitalna srčana mana. **Prikaz slučaja.** Prikazali smo mladića starog 20 godina, aktivnog sportistu, kik boksera, koji je upućen u Centar za kardiologiju Kliničkog centra Crne Gore, u cilju redovne provere kardiološkog statusa. Neposredno nakon rođenja bolesnik je operisan zbog traheoezofagealne fistule. Rutinski ehokardiografski pregled u četvrtoj godini života pokazao je do tada neprepoznati DOMV, bez prisustva stenoze ili regurgitacije i bez drugih anomalija na srcu. Na pregledu kod kardiologa za odrasle, u 20. godini života mladić je bio bez ikakvih tegoba. Objektivnim pregledom nalaz na srcu bio je uredan. Elektrokardiografom registrovan je sinusni ritam, nekompletni blok desne grane. Na poprečnom transtoraksnom ehokardiografskom pregledu viđena su dva asimetrično postavljena otvora bez prisutne regurgitacije ili stenoze. Ostali nalaz bio je uredan. Transezofagusnim ehokardiografskim (TEE) pregledom u nivou mitralne valvule ustanovljena su 2 asimetrična mitralna orificijuma odvojena fibroznim tkivom, mitralni kuspisi uz separatnu inserciju hordi za svaki orificijum. **Zaključak.** Prikazani bolesnik je jedini dijagnostikovani bolesnik u Crnoj Gori. Posebno je značajna činjenica da za vreme praćenja od 16 godina i aktivnog bavljenja vrlo zahevnim sportom nisu nastale funkcionalne promene na srcu. Takođe, ne postoje preproruke u literaturi za ovaj problem.

Ključne reči:
srce, kongenitalne mane, dijagnoza; ehokardiografija, transezofagusna.

Introduction

Double orifice mitral valve (DOMV) is a very rare congenital heart defect. Greenfield [1] was the first author who described this heart defect in 1876. Until today about 200 cases of double orifice mitral valve were described [2]. Approximately 50% cases of double orifice mitral valve are detected during investigation of other congenital heart disease [2].

Mitral valve consists of a central orifice located between the sail-like anterior leaflet and a C-shaped posterior

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mitral leaflet. The anterior cusp occupies roughly one third of the annular circumference and the posterior leaflet occupies about the remaining two-thirds of the annular circumference [3]. DOMV is a defect with two separate mitral orifices separated by fibrous tissue [2]. Pathoanatomic substrate are two openings. Usually in 85% of cases the eccentric orifice located on anterolateral or posteromedial commissure. In the minority of cases there is a central type and fibrous tissue that separates the two symmetrical holes [3]. In DOMV the mitral valve is functioning normally in about 50% cases and in the other 50% cases the valve is functioning like stenotic or there is regurgitation [4]. Some authors describe a duplicate of the mitral valve: this condition involves two mitral valves and each of them with its own set of leaflets, commissures, chordae and papillary muscles [5]. Double orifice mitral valve was found in 1% of autopsied cases of congenital heart disease in the Cardiac Registry of the Children's Hospital, Boston [6]. Postmortem, in 27 cases an anomaly of the tensor apparatus was always found [6]. These malformations may be associated with the chordal ring, accessory papillary muscle or muscles, subdividing muscular ridge, fused papillary muscles (parachute mitral valve), crossing chordae tendines, and central fibrous subdivision [6]. The embryologic theories explaining this abnormal leaflet fusion and persistence of the left part of the common atrioventricular canal. Abnormal structure, including large bridging tissue, abnormal leaflets, chordae fused or abnormal papillary muscles, reduce the effective area and valves can result in clinically significant degrees of mitral incompetence [7, 8]. DOMV without mitral regurgitation or mitral stenosis is asymptomatic [9]. Surgical intervention is necessary when stenosis or incompetence is severe or if repair of an associated cardiac lesion is needed [9].

A total of 46 children, aged 2 days to 16 years with DOMV were studied and partial atrioventricular septal defect was the most commonly associated cardiac lesion [10]. Symptoms were related to the degree of mitral insufficiency and/or stenosis when present. Surgical intervention directed at double orifice mitral valve was required in the minority of patients who underwent repair of associated cardiac lesions. The long-term morbidity attributable to DOMV was low [10]. Other associated lesions include atrial septal defects (ASD), ventricle septal defect (VSD), coarctation of the aorta, tetralogy of Fallot, patent ductus arteriosus, hypoplastic left heart syndrome, left superior vena cava, unroofed coronary sinus, Ebstein anomaly of the tricuspid valve, dysplastic tricuspid valve, double-orifice tricuspid valve, Shone syndrome, parachute mitral valve, flail mitral-valve leaflet, truncus arteriosus, pulmonary stenosis, bicuspid aortic valve, left ventricular noncompaction (isolated and nonisolated types), tricuspid atresia, and corrected transposition [10–19].

Case report

The presented 19-year-old patient male is the only one patient registered in our country, which has 650,000 inhabitants. The congenital heart disease, DOMV, was accidentally discovered 4 years after the operation due to tracheoesophageal fistula. It was the isolated form without the presence of mitral stenosis and mitral regurgitation, with the normal dimensions of all the cardiac chambers. At the age of 19 years, he was examined by the adult cardiologist. He had no complaints. He was an active athlete − kick boxer. Physical finding was normal, ECG showed regular sinus rhythm with incomplete right bundle branch block. Chest X ray demonstarted the normal size of the heart. Transthoracic 2D and 3D echocardiography was perfomed with a Phillips HD11XE ultrasound machine.

Transthoracic echocardiography (TTE) examination registered in sectional parasternal long axis view: normal left atrium size, mitral leaflets slightly more voluminous, the left ventricle of completely normal size without segment changes into kinetic with preserved systolic function. The root of the aorta was of normal width, aortic valve with three cusps, leaflet thin with good separation. The ascending aorta was of completely normal width (Figure 1). On sectional parasternal short axis view there was a double mitral orifice, mitral valve was divided into two separate valve orifices (anterolateral and posteromedial) by a fibrous bridge through which registered normal flow (Figure 2).

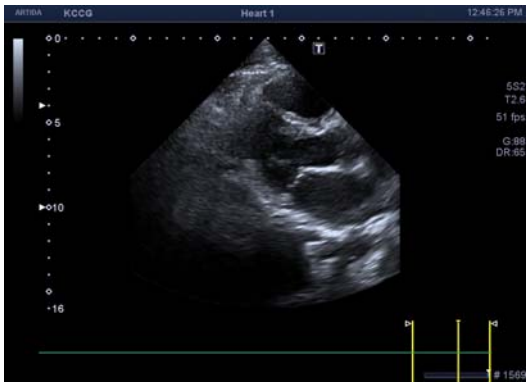

Fig. 1 – Long axis view – without abnormalities.

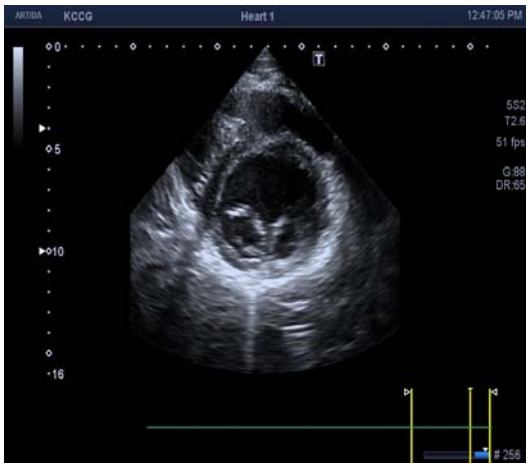

Fig. 2 – Short axis view – double mitral valve orifice.

On the apical 4-chamber view, the central part of the anterior mitral leaflet, the fibrous bridge did not move toward the apex at diastole while the medial and lateral parts did (Figure 3). Four-chamber view of the tricuspid valve and pulmonary artery valve looked normal. A trace of tricuspid regurgitation

and systolic pressure were registered in the right ventricle (RVSP) of 20 mmHg. Right cavities were completely normal. The pericardium was normal (Figure 3). Transesophageal echocardiography showed the transgastric review two separate asymmetric mitral orifice separated by fibrous tissue, mitral leaflet with a separate one for each insertion hordes orifice (Figure 4). Blood flow through both the orifices was normal. No other congenital cardiac anomaly was detected.

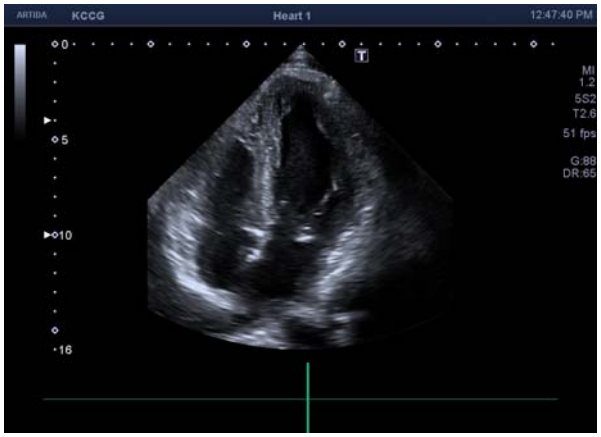

Fig. 3 – Four chamber view − unusual view of the mitral valve during systole; center part of the anterior mitral leaflet, the fibrous bridge did not move toward the apex at diastole while the medial and lateral parts did.

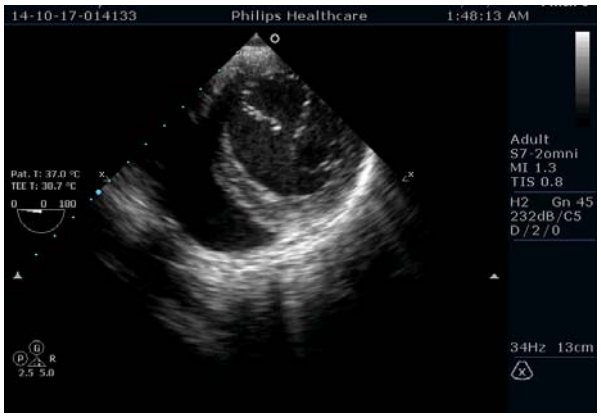

Fig. 4 – Transesophageal echocardiography (TEE) transgastric view – unequal holes in the double orifice mitral valve.

Discussion

DOMV is a rare congenital heart disease. Based on echocardiography recording Trowitzsch et al. [5] described 3 types of DOMV: hole type (accessory orifice surrounded by leaflet tissue that may have a chordal ring), complete bridging (fibrous bridge in the plane of the mitral valve sails, dividing the mitral valve opening into two parts that may be equal or unequal), and incomplete bridging − a small strand of fibrous tissue connects only the tips of the anterior leaflets separator insertion hordes of every orifice. The presented patient had unequal, asymmetric position of orifices.

The mitral valve is functionally normal in about 50% of patients and significant regurgitation or stenosis is found in the remaining patients [8]. In a published clinical series of 46 children with DOMV, the authors reported mitral regurgitation as the most frequent association, detected by color Doppler in 43% of the patients [10]. DOMV with normal flow was seen in 37% of the patients, and mitral stenosis in 13% of the patients [10]. Physical findings may only exist if mitral regurgitation or mitral stenosis are present [20]. The presented patient had an isolated form without the presence of mitral stenosis and mitral regurgitation, so physical finding was normal.

Associated congenital heart defects are common, although DOMV can occur as an isolated anomaly. The most common associated lesion is atrioventricular septal defect found in 4.9−17.9% of cases in necropsy studies [5]. The presented pateint had no associated anomalies.

Double orifice mitral valve has also been reported in patients presenting with atrial tachycardia or congenital complete heart block [18, 19]. The presented patient had no rhythm disturbance. The problem is that there is no recommendation for patients with DOMV could they be actively engaged in sport. Physical activity restriction is recommended for symptomatic patients [21].

Treatment depends on the type and severity of mitral valve dysfunction. Isolated DOMV causing neither obstruction nor regurgitation needs no active intervention [9]. Long-term follow-up is necessary to detect abnormal hemodynamics or subsequent complications [22]. Mitral stenosis and regurgitation were found together in 6.5% of the patients, but only 16% of the patients had severe enough to require surgical lesions intervention [10]. According to the updated guidelines of the American Heart Association, patients with double orifice mitral valve do not need antibiotic prophylaxis against infective endocarditis [21]. Patients with DOMV with mitral stenosis are amenable to percutaneous transcatheter balloon dilatation [23]. Surgical repair should be performed in all symptomatic patients with DOMV which has regurgitation and should also be performed in patients with stenotic DOMV, if balloon dilation is not feasible [21]. Follow-up care is necessary for prompt detection and treatment of infections, arrhythmia and other complications [21].

Conclusion

The presented patient with DOMV is the only one recognized in our country. It is the isolate form of this rare disease, asymptomatic, without associated anomalies, as well as without mitral regurgitation and mitral stenosis. The case is interesting because during a 16-year follow-up period there were no functional changes despite the fact that he performed a very demanding sport activity. This is highly important because there is no information in the literature about that.

Declaration of interest

All the authors declare no conflict of interest.

R E F E R E N C E S

1. *Greenfield WS*. Double mitral valve. Trans Pathol Soc (London) 1876; 27: 128−9.
2. *Anwar AM, McGhie JS, Meijboom FJ, Ten CF*. Double orifice mitral valve by real-time three-dimensional echocardiography. Eur J Echocardiogr 2008; 9(5): 731−2.
3. *Tandon R, Takkar S, Kumbhkarni S, Kumar N, Aslam N, Mohan B,* et al. A rare case of double orifice mitral valve with perimembranous ventricular septal defect: Application of three-dimensional echocardiography for clinical decision making. Ann Pediatr Cardiol 2010; 3(1): 87−9.
4. *Ozeke O, Ozbakir C, Gunel EN*. Double Mitral Valve Imaging. J Am Soc Echocardiogr 2010; 23(3): 340.e1−2.
5. *Trowitzsch E, Bano-Rodrigo A, Burger BM, Colan SD, Sanders SP*. Two-dimensional echocardiographic findings in double orifice mitral valve. J Am Coll Cardiol 1985; 6(2): 383−7.
6. *Baño-Rodrigo A, van Praagh S, Trowitzsch E, van Praagh R*. Double-orifice mitral valve: A study of 27 postmortem cases with developmental, diagnostic and surgical considerations. Am J Cardiol 1988; 61(1): 152−60.
7. *Hoffman P, Stümper O, Groundstroem K, Sutherland G*. The transesophageal echocardiographic features of double-orifice left atrioventricular valve. J Am Soc Echocardiogr 1993; 6(1): 94−100.
8. *Karas S, Barbetseas J, Lambrou S, Parissis J, Metzikof D, Toutouzas P*. Well-functioning double-orifice mitral valve in a young adult. J Clin Ultrasound 2003; 31(3): 170−3.
9. *Taksande A, Goutami V, Thomas E*. Double orifice mitral valve associated with Ventricular Septal Defect in an infant: Case Report. Pediatric Cardiol 2011; 13(1): 6−9.
10. *Zalzstein E, Hamilton R, Zucker N, Levitas A, Gross GJ*. Presentation, natural history, and outcome in children and adolescents with double orifice mitral valve. Am J Cardiol 2004; 93(8): 1067−9.
11. *Yesilbursa D, Miller A, Nanda NC, Mukhtar O, Huang WY, Ansingkar K,* et al. Echocardiographic diagnosis of a stenotic double orifice parachute mitral valve with a single papillary muscle. Echocardiography 2000; 17(4): 349−52.
12. *Sugiyama H, Hoshiai M, Toda T, Nakazawa S*. Double-Orifice Mitral Valve Associated with Noncompaction of Left Ventricular Myocardium. Pediatr Cardiol 2006; 27(6): 746−9.
13. *Shiran A, Lewis BS*. Acquired partially flail leaflet causing severe mitral regurgitation in a congenital double-orifice mitral valve. J Am Soc Echocardiogr 2004; 17(5): 478−9.
14. *Kim S, Shin E, Lee S*. Congenital double-orifice mitral valve with mitral regurgitation due to flail leaflet in an elderly patient. Korean J Intern Med 2005; 20(3): 251−4.
15. *Gerber IL, Calder LA, Ruygrok PN, Greaves SC, Gibbs HC, Long S,* et al. Association of a double orifice mitral valve with a bicuspid aortic valve in an explanted heart with dilated cardiomyopathy. Heart Lung Circ 2003; 12(3): 188.
16. *Park IS, Yoo SJ, Kim KS, Hong CY*. Tricuspid atresia associated with double-orifice mitral valve and coronary sinus septal defect. Tex Heart Inst J 1991; 18(3): 202−5.
17. *Abdullah M, Pearce K, Palmer N, Chenzbraun A*. Double orifice mitral valve with dysplastic tricuspid valve and intact interatrial septum: a three-dimensional echocardiographic study. Eur J Echocardiogr 2008; 9(4): 598−9.
18. *Westendorp IC, de Bruin-Bon HA, Hrudova J*. Double orifice mitral valve; a coincidental finding. Eur J Echocardiogr 2006; 7(6): 463−4.
19. *Warnes C, Somerville J*. Double mitral valve orifice in atrioventricular defects. Br Heart J 1983; 49(1): 59−64.
20. *Lu Q, Lu X, Xie M, Wang X, Wang J, Yang Y,* et al. Real-time three-dimensional echocardiography in assessment of congenital double orifice mitral valve. J Huazhong Univ Sci Technol Med Sci 2006; 26(5): 625−8.
21. *McElhinney DB, Sherwood MC, Keane JF, del Nido PJ, Almond CS, Lock JE*. Current management of severe congenital mitral stenosis: outcomes of transcatheter and surgical therapy in 108 infants and children. Circulation 2005; 112(5): 707−14.
22. *Hartas GA, Rao PS, Balaguru D*. Mitral Valve, Double Orifice. eMedicine from WebMD. Available from: http://emedicine.medscape.com/article/897322-overview. [updated 2014 October 05].
23. *Kim MH, Cha KS, Kim JS, Hung JS, Lau KW*. Successful inoue balloon mitral commissurotomy in double-orifice mitral stenosis. Catheter Cardiovasc Interv 2000; 49(2): 200−3.

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