Congenital double-orifice mitral valve associated with atrioventricular septal defect: A case report

Osman Küçükosmanoğlu[1], Nazan Özbarlas[1], Orhan Kemal Salih[2]

Departments of [1]Pediatric Cardiology, and [2]Cardiovascular Surgery, Çukurova University Faculty of Medicine, Adana, Turkey

SUMMARY: Küçükosmanoğlu O, Özbarlas N, Salih OK. Congenital double-orifice mitral valve associated with atrioventricular septal defect: a case report. Turk J Pediatr 2002; 44: 160-161.

The case of an eight-year-old girl with congenital double-orifice mitral valve associated with atrioventricular septal defect is reported. This rare mitral valve anomaly was diagnosed with two-dimensional echocardiography. The associated atrioventricular septal defect was repaired successfully. The central fibrous bridge between anterior and posterior mitral valve leaflets was left undivided to avoid iatrogenic mitral regurgitation. Preoperative diagnosis may provide some useful information to the surgeon. The surgical method can be individualized in each case.

Key words: double-orifice mitral valve, atrioventricular septal defect, echocardiography.

Double-orifice mitral valve (DOMV) is a rare condition and may be associated with other cardiac anomalies such as subaortic ring, coarctation of aorta, patent ductus arteriosus, pulmonary stenosis, ventricular septal defect, truncus arteriosus and primum type atrial septal defect[1-6]. The orifices are usually unbalanced in size and there is a central fibrous bridge between the anterior and posterior mitral valve leaflets. The medial and lateral orifices are equal in size in only 15% of patients[1].

Herein, we report an eight-year-old girl with balanced DOMV and primum type atrial septal defect who was diagnosed with two-dimensional echocardiography and was successfully treated with surgical intervention.

Case Report

An eight-year-old girl was referred to our department for echocardiographic study because of heart murmur. She had growth retardation with stature of 109 cm (3rd percentile), and weight of 17 kg (<3rd percentile). Physical examination revealed normal blood pressure, regular pulse of 88 bpm, fixed splitting of second heart sound at the upper left sternal edge and 3rd degree apical pansystolic murmur. The other findings were all normal. Chest X-ray showed marked cardiomegaly with a cardiothoracic index of 0.64 and increased vascularization. Echocardiogram (ECG) showed normal sinus rhythm, left QRS axis (-45°) deviation and incomplete right bundle-branch block. Two-dimensional echocardiography showed a large primum type atrial septal defect, small mitral cleft and balanced DOMV (Fig. 1), and color Doppler showed moderate mitral regurgitation. Cardiac catheterization revealed mild pulmonary hypertension with a mean pulmonary artery pressure of 27 mmHg and increased pulmonary flow with a Qp/Qs ratio of 3.2/1.

Fig. 1. Two-dimensional echocardiographic view of double-orifice mitral valve in parasternal short-axis window.

The primum atrial septal defect was repaired with pericardial patch without complication. The cleft was partially sutured to avoid creating mitral stenosis and the bridging tissue was left undivided to prevent more mitral regurgitation. After one year follow-up she caught up growth with a stature of 119 cm and a weight of 22 kg. Chest X-ray showed a normal cardiothoracic index and echo-Doppler study showed trivial mitral regurgitation.

Discussion

Congenital mitral valve anomalies are rare conditions and are usually associated with some other cardiac defects. Banerjee et al.[3] reported 65 children with congenital mitral valve anomalies in their study, which consisted of 13,400 new echocardiographic data during 7.5 years, and only seven of the children had DOMV.

As well as in other mitral valve anomalies, two-dimensional echocardiography is an extremely useful method for the diagnosis of DOMV. Two separate holes in the mitral valve can be identified in parasternal short-axis view. The color Doppler echocardiography provides an adequate anatomical and functional assessment of DOMV[3-4].

The diagnosis of DOMV prior to surgical intervention for associated defects is very important. If the central fibrous bridge between the anterior and posterior mitral valve leaflets causes functional mitral stenosis, it may require resection. But in patients without functional mitral stenosis it may be beneficial not to incise the bridging tissue to avoid iatrogenic mitral regurgitation[7]. Valve replacement should be the treatment of choice in severe mitral regurgitation with DOMV. Yurdakul et al.[8] reported successful valve replacement in an 18-month-old boy with severe mitral regurgitation associated with DOMV.

The DOMV may be an incidental echocardiographic finding but it is usually associated with another cardiac anomaly. Preoperative echocardiographic evaluation of the mitral valve may provide some useful information to the surgeon. The surgical method can be indivualized in each case.

REFERENCES

1. Baylen BG, Gleason MM, Waldhausen JA. Mitral inflow obstruction. In: Emmanouilides GC, Reimenschneider TA, Allen HD, Gutgesell HP (eds). Heart Disease in Infants, Children, and Adolescents (5th ed) Vol 2. Baltimore:Williams and Wilkins; 1995: 1042-1054.
2. Yurdakul Y, Arsan S, Karapınar K, Tamim M, Bilgiç A. Congenital double-orifice mitral valve: a case report. Turk J Pediatr 1995; 37: 173-176.
3. Banerjee A, Kohl T, Silverman NH. Echocardiographic evaluation of congenital mitral valve anomalies in children. Am J Cardiol 1995; 76: 1284-1291.
4. Solorio S, Badui E, Yanez M, Encisio R, Rodriguez L, Quintero LR. Double mitral valve orifice. Two dimensional and Doppler echocardiographic diagnosis. Arch Med Res 1996; 27: 491-494.
5. Anzai N, Yamada M, Tsuchida K, Yoshioka Y, Iida Y, Wakabayashi Y. Double orifice mitral valve associated with endocardial cushion defect. Jpn Circ J 1986; 50: 455-458.
6. Silverman NH. Pediatric Echocardiography. Baltimore: Williams and Wilkins; 1993: 239.
7. Bono-Rodigo A, Van Praagh S, Trowitzsch E, et al. Double orifice mtral valve: a study of 27 postmortem cases with developmental, diagnostic and surgical considerations. Am J Cardiol 1988; 61: 152-160.
8. Yurdakul Y, Bilgiç A, Saylam A, Sarıoğlu T, Koşker S, Aytaç A. Congenital double-orifice mitral valve. report of a case with valve replacement. Jpn Heart J 1980; 21: 545-550.