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Double Orifice Mitral Valve Associated with Ostium Primum Atrial Septal Defect

(Case Report)

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The double orifice mitral valve was first described by Greenfield in 1966 and has been reported in over 40 patients to date (quoted by Mercer and Tubbs).[1] Double orifice mitral valve, or "duplication of the mitral valve" has also been observed in some animals, e.g. calf and yak.[2] This rare anomaly usually presents a congenital mitral stenosis or may show normal valve function.[1,2] Mitral regurgitation has been reported in only one case, combined with subaortic stenosis.[1] We would like to present a case of double orifice valve with ostium primum atrial septal defect, where the mitral valve showed mild insufficiency.

T.K. was a 20 year old female patient who was admitted to the Hacettepe Children's Hospital, Department of Thoracic and Cardiovascular Surgery, with the complaints of dyspnea and palpitation. Physical findings at the time of admission revealed blood pressure to be 90/50 mm/Hg and pulse rate 76/min; a 4/6 pansystolic murmur, heard best in the intercostal space along the left sternal border, and a 2/6 systolic murmur at the apex were present. The liver was palpable 3 cm below the right costal margin.

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ECG findings were compatable with an incomplete right bundle block; chest X-ray displayed left ventricular hypertrophy.

This patient's past history revealed open heart surgery in 1963 when a primum atrial septal defect was closed with primary sutures without teflon patch; no other pathology was suspected during her first operation.

Cardiac catheterization and angiography were performed which showed a left to right shunt at atrial level and mild mitral regurgitation. Values obtained from right and left heart catheterizations are tabulated in Table I. A diagnosis of atrial septal defect and mild mitral insufficiency was made, and the patient underwent cardiopulmonary bypass with Rygg-Kyvsgaard disposable bubble oxygenator, DeBakey roller pumps; was instituted after a sternal split was performed and pericardiomyocardial adhesions due to the first operation were freed. The patient was cooled to 32 °C. Right atriotomy was done after cross-clamping of the aorta. A primum type atrial septal defect, 3 cm in diameter was observed. The mitral valve was duplicated. Posterior orifice was slightly smaller than the anterior orifice. The atrial septal defect was closed with a teflon patch by continuous silk sutures. No surgical intervention was thought to be necessary for the duplicated mitral valve because combining the two mitral orifices by cutting the middle bar would have resulted in a very large mitral annulus not suitable for even the largest prosthetic valve.

TABLE I

CATHETERIZATION DATA

	Oxygen (%)	Pressures (mmHg)	
RIGHT HEART:			
Pulmonary capillary			15 (mean)
Pulmonary artery	85	35/25	25 (mean)
Right ventricle	80	32/12	
Right Atrium:			
Upper level	63		
Middle level	80		13 (mean)
Lower level	70		
Superior vena cava	58		
Inferior vena cava	60		
LEFT HEART:			
Femoral artery	98		
Left ventricle	—	116/15	
Aorta	—	116/75	90 (mean)

Pulmonary/Systemic flow ratio: 3

The postoperative course of the patient was uneventful; she did not require to be digitalized and was discharged from hospital on the 12th postoperative day with a soft systolic murmur (grade II/VI) in good condition.

Discussion

Duplication and triplication of the mitral valve is very rarely encountered as congenital deformities, duplication being more common than triplication.[3] Schraft and Lisa[4] stated that duplication of the mitral valve (double orifice mitral valve) might be the result of foetal endocarditis or may be a purely developmental anomaly. The fact that duplication of the mitral valve is associated with partial atrioventricular canal of 25 percent of the cases, strongly suggests that this rare anomaly is related to the development of endocardial cushions.[2] Two, three or four papillary muscles may be present in the left ventricular cavity and each orifice of the mitral valve may have separate chordae tendineae.[2] The mitral orifices may be either equal or unequal in size.[2] It was observed by Wigle,[3] that combined areas of the double orifices may be less than the normal mitral area. One of the orifices in duplicated mitral valve is sometimes incompetent and this part of the valve receives accessory chordae and papillary muscles from the subaortic area, which might be responsible for mitral insufficiency as was thought in Mercer and Tubbs'case.[1]

Duplication of the mitral valve may occur as an isolated lesion and it is associated with other cardiac pathologies in 54 % of the cases, among which endocardial cushion defects are most commonly encountered.[2] Pyorala and co-workers (cited by Mercer and Tubbs)[1] listed the associated defects in order of frequency as follows: Persistent atrioventricular canal (partial or complete), bicuspid aortic valve, coarctation of the aorta, patent ductus arteriosis, right sided aortic arch, subaortic stenosis, bicuspid pulmonary valve, Ebstein's anomaly and secundum atrial septal defects.

Double orifice mitral valve can sometimes be overlooked during operations for atrial septal defects. Such a case was reported by Reed et al.[5] This was a child who had undergone closure of an atrial septal defect, 2 years before he had a fatal accident. Autopsy of the child revealed duplication of the mitral valve and bicuspid aortic valve, although no pathology except the atrial septal defect was noted during his operation. The presence of double orifice mitral valve was also overlooked during the first operation of our patient. Cases with primum type atrial

septal defects should be examined carefully for the presence of duplicated mitral valve, as 5 % of the patients with partial persistent atrioventricular canal have double orifice mitral valve.[2]

No surgical intervention was reported until Reed and co-workers case, and older cases were presented as incidental findings at portmortem examinations.[4, 5, 6] Reed et al[5] divided the bridge of tissue between two mitral orifices and obtained a good surgical result in their case, which also had on ostium primum atrial septal defect. A hemodynamically insignificant soft systolic apical murmur was heard after the operation. Mercer and Tubbs[1] presented a case of duplicated mitral valve associated with subaortic stenosis and closed the smaller mitral orifice with interrupted mattress sutures and teflon pledgets. Pernot[7] reported a case of double orifice mitral valve who had undergone mitral valve replacement.

In our case no surgical intervention was done because of the presence of hemodynamically insignificant mitral insufficiency. It may be difficult to decide which of the orifices is incompetent. Closure of the smaller orifice may result in postoperative mitral stenosis. We believe that mitral valve replacement should be the treatment of choice where mitral regurgitation is hemodynamically important, and when closure of the smaller orifice is not compatible with hemodynamically normal valve function. In cases of mitral stenosis associated with duplicated mitral valves, cutting the bridge of tissue separating the two mitral orifice should be tried first.

Summary

A case of primum type atrial septal defect and double orifice mitral valve, where no surgical intervention was done for the duplicated mitral valve, because of the hemodynamically insignificant mitral regurgitation is presented. Literature concerning this rare pathology is reviewed and it was concluded that mitral valve replacement should be the treatment of choice if mitral regurgitation is important hemodynamically and closure of the smaller orifice is not compatible with normal mitral valve function. Cutting the central bar between the two mitral orifices should be tried first in cases of mitral stenosis. It might be difficult to decide which of the two orifices is incompetent and closure of one of them during the operation may result in postoperative mitral stenosis or insufficiency. Careful examination of the mitral valve is mandatory particularly in cases of partial or complete atrioventricular canals, taking into consideration that this rare malformation of the mitral valve is usually associated with endocardial cushion defects.

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