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CONGENITAL DOUBLE-ORIFICE MITRAL VALVE*
A Case Report

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SUMMARY: Yurdakul Y, Arsan S, Karapınar K, Tamim M, Bilgiç A. (Department of Thoracic and Cardiovascular Surgery, Hacettepe University Faculty of Medicine, Ankara, Turkey). Congenital double-orifice mitral valve. Turk J Pediatr 1995; 37: 173-176.

A three-year-old girl was admitted to Hacettepe Children's Hospital due to sweating and poor feeding. Echocardiography demonstrated ostium primum atrial septal defect with a cleft mitral valve. After two months of medical follow-up with digitalis, she was subjected to open-heart surgery, and a primum-type atrial septal defect with double mitral orifice was found. To avoid creating acute mitral stenosis, the cleft was only partially sutured and the septal defect was closed with a patch. Her postoperative recovery was uneventful. It is emphasized that surgical intervention should be individualized in each case. *Key words: congenital heart defect, congenital double-orifice mitral valve, atrial septal defect.*

Duplication of the mitral valve is an extremely rare congenital cardiac malformation. In half of the reported cases, the duplication of the mitral valve occurs as an isolated lesion[1]. Atrioventricular septal defect appears to be the most common among the associated cardiovascular anomalies.

This malformation of the valve sometimes presents with no hemodynamic disturbances, but usually produces mitral stenosis and, very rarely, mitral insufficiency[1,2].

A case of duplication of the mitral valve for which the preoperative diagnosis was primum-type interatrial septal defect with a cleft mitral valve is the subject of this paper.

Case Report

A three-year-old girl was admitted to Hacettepe Children's Hospital in December 1992 due to sweating and poor feeding. Physical examination revealed a heart

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rate of 112/min in sinus rhythm and blood pressure of 110/60 mmHg. Precordial thrill, pansystolic murmur, early diastolic murmur, and an ejection systolic murmur in the pulmonary area were noted. Further examination was unremarkable. The electrocardiogram showed left-axis deviation and left ventricular hypertrophy.. Chest x-ray revealed global cardiac enlargement with pulmonary plethora (Fig. 1). Echocardiography demonstrated ostium primum atrial septal defect with a cleft mitral valve. The left ventricular angiogram revealed the characteristic "goose neck" deformity of the left ventricular outflow tract and mitral insufficiency. In cardiac catheterization, the pulmonary-to-systemic flow ratio was 3.9 at the atrial level, the pulmonary artery pressure was 32/6 and the mean left atrial pressure was 6 mmHg.

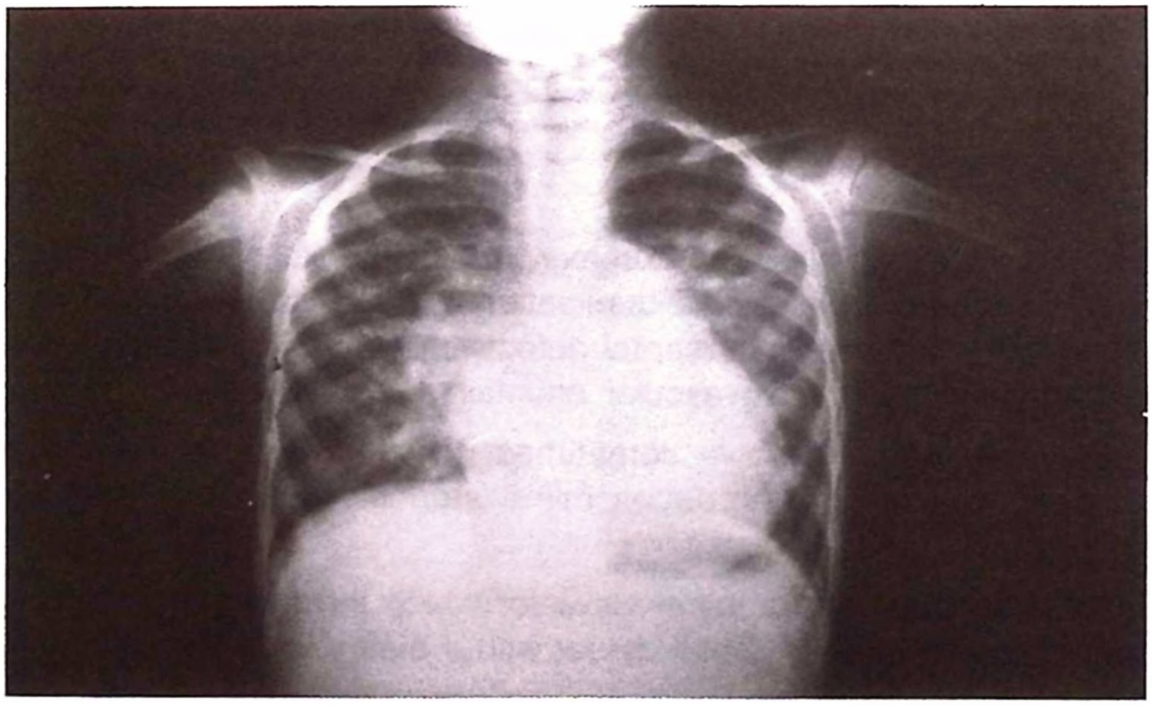

Fig. 1: Chest roentgenogram demostrating cardiomegaly and increased pulmonary vascularity.

After two months of medical follow-up with digitalis, the patient underwent open-heart surgery using the Bentley bubble-oxygenator and DeBakey roller pumps under hypothermia of 25 °C. Before cardiopulmonary by-pass was started, the digital examination through the right atrial appendix verified the presence of mitral insufficiency. Intraoperatively, an ostium primum-type atrial septal defect 1.5 cm in diameter was seen. The mitral valve had a double orifice-one side was 1 cm in diameter, located posteromedially, and the other 2.5 cm in diameter, located anterolaterally with cleft towards its anterior aspect. The double orifice was separated by a fibrotic band, and there were two underlying papillary muscles supplying chordae to both cusps of the appropriate orifice; however, for the area adjacent to the cleft, chordal support arose directly from the ventricular wall. To

avoid creating acute mitral stenosis, the cleft was only partially sutured, and the posteromedially located orifice was left untouched. To close the septal defect, a dacron patch was affixed with interrupted sutures to the ventricular septum, the tricuspid valve and the right atrial wall anterior and inferior to the coronary sinus orifice, and continuous sutures were used along the remainder of the defect.

The patient's postoperative recovery was uneventful, and she was kept on a daily dose of digitalis but rarely on diuretics. Postoperative echocardiography demonstrated minimal mitral insufficiency and double orifice of the mitral valve (Fig. 2).

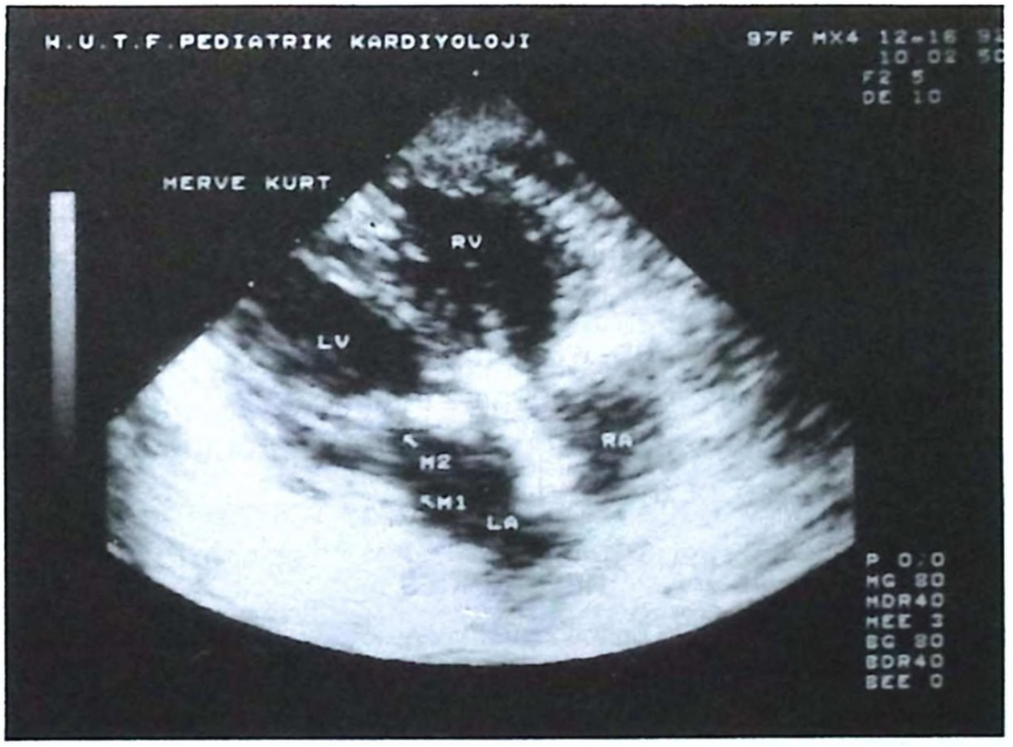

Fig. 2: Postoperative echocardiography showing double orifice of the mitral valve.

Discussion

In 25 percent of the reported cases, partial atrioventricular septal defect is associated with this rare malformation, suggesting that the developmental defect involves endocardial cushion tissue. Since more than two left ventricular papillary muscles are associated with malformation, it is now accepted that the true developmental basis of duplication of the mitral valve is unknown[1]. In this malformation, the two mitral orifices are usually of unequal size, and additional papillary muscles and chordae tendineae may be present[1]. There had been no surgical approach reported until Reed et al's[4] presentation. Since then surgical management has improved tremendously, parallelling the better understanding of the pathology. A careful appraisal of the structure of the valve (site of the accessory orifice, cleft leaflet or a parachute valve) and associated malformations

may guide the surgeons to decide whether any surgical intervention on the valve is warranted[2]. Duplication of the mitral valve may function normally, requiring no repair. In selected cases conservative management may be preferable[2,3].

There are three types of surgical approach: major valve reconstruction, cleft suture, and valve replacement[2,4-8]. Major valve reconstruction by the Amano-Suzuki technique, i.e. division of the fibrous bar, valve reconstruction by cleft suture and leaflet extension may be considered by experienced surgeons when there is a failure of leaflet coaptation due to annular dilatation or leaflet retraction[5]. In some cases the fibrous bar may contain leaflet tissue responsible for keeping the valve competent. If the valve is incompetent due to a cleft leaflet, the cleft may be sutured. Partially cleft suture may be preferred over complete cleft suture because of a lower risk of acute mitral stenosis. However, the possibility of the development of late mitral stenosis still exists. If one orifice is incompetent, its closure may be considered[2]. If the valve is severely damaged causing significant mitral insufficiency or stenosis, valve replacement should be advocated[7,9]. We previously published a case of an 18-month-old child with a double orifice mitral valve who was successfully treated with mitral valve replacement because of severe mitral valve insufficiency[7,9]. Care must be taken to avoid creating iatrogenic valve stenosis by suturing of the cleft or acute mitral incompetence by division of the bridge[2].

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