Congenital Double-Orifice Mitral Valve

Report of a Case With Valve Replacement

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SUMMARY

A case of double-orifice mitral valve (duplication of the mitral valve) in an 18-month-old baby is presented. The valve showed significant regurgitation and was replaced with a No. 20 Lillehei-Kaster mitral prosthesis. Patient is kept on dipyridamole (Persantin) 10 mg/Kg and aspirin 50 mg/Kg daily to avoid postoperative thromboembolic complications.

It is emphasized that valve replacement should be the treatment of choice in severe mitral regurgitation associated with double-orifice mitral valve.

Additional Indexing Words:

Congenital mitral regurgitation Congenital mitral stenosis Valve replacement in children Thromboembolism Dipyridamole (Persantin) Aspirin

DOUBLE-ORIFICE mitral valve is an extremely rare congenital cardiac malformation and has been observed in over 40 patients to date (quoted by Mercer and Tubbs).[1] Carpentier et al[2] has encountered no case of double-orifice mitral valve in 47 children with congenital mitral valvulopathies in a cooperative study from 3 major cardiac centers in Europe.

There had been no surgical approach for this pathology until the publication by Reed et al,[3] and this malformation has been reported as an incidental postmortem finding in most of the cases since the original description of this

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pathology by Greenfield in 1876 (cited by Mercer and Tubbs).[1] This malformation usually presents itself as mitral stenosis or sometimes shows normal valve function. Mitral regurgitation has been very infrequently reported in this lesion.[1),4),5)]

A case of double-orifice mitral valve with severe mitral regurgitation, treated with valve replacement, is the subject of this paper. This is the second case of double-orifice mitral valve encountered among 18 cases of congenital mitral valvulopathies in our clinic, where no surgical intervention was performed in the first one owing to the insignificant mitral regurgitation, which was associated with ostium primum atrial septal defect.[5)]

Case Report

E.E. (File No. 896224), an 18-month-old male baby, admitted to our hospital due to frequent attacks of upper respiratory tract infection. He was an underdeveloped child weighing 8,500 Gm. Cyanosis of the fingertips and the mouth, and engorged cervical veins were observed on inspection.

Physical examination revealed blood pressure 100/70 mmHg and pulse rate 144/min in sinus rhythm. Precordial thrill, apical holosystolic murmur and crepitations in the lung fields were noted and the liver was palpable 3 cm below the costal margin. Examination of the other systems was non-contributory.

Chest X-ray revealed global cardiac enlargement and increased pulmonary vascularity (Fig. 1). ECG displayed left atrial dilatation and left ventricular hypertrophy. Contrast material injected into the left ventricle during angiography proved the presence of severe mitral regurgitation (Fig. 2).

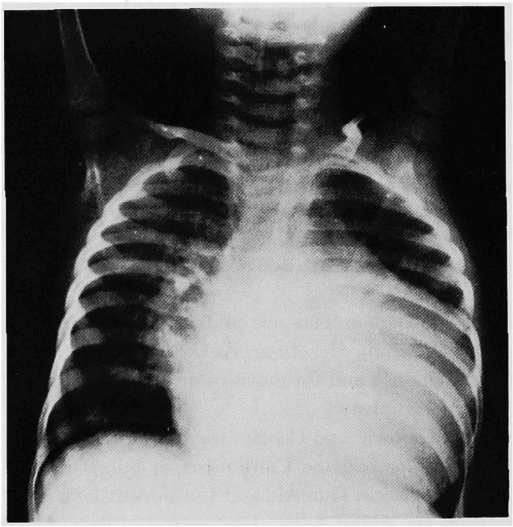


Fig. 1. Preoperative chest X-ray showing global enlargement of the cardiac silhouette.

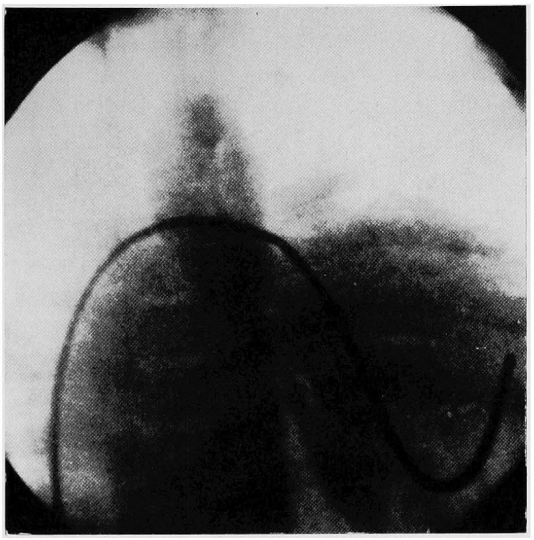
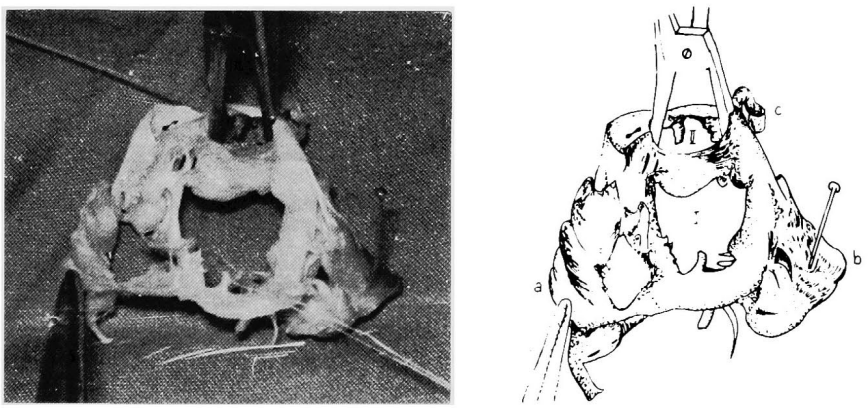


Fig. 2. Preoperative left ventriculography displaying significant degree of mitral regurgitation. Note the enlarged left atrium filled with the contrast material injected into the left ventricle.



(A) (B)

Fig. 3. A) Picture of the resected specimen. Double orifice mitral valve is shown.

B) Schematic drawing of the resected specimen shown in Fig. 3A. I = larger orifice, 3.5 cm in diameter; II = smaller orifice, 1 cm in diameter; a, b, and c = remnants of papillary muscles and chordae tendineae on the resected specimen.

Patient was given digitalis and diuretics during the preoperative period. He was subjected to open heart surgery on January 1, 1978, using Bentley bubble-oxygenator and DeBakey roller pumps, under hypothermia of 25°C. Left atrium was opened after aortic cross-clamp. Mitral valve showed double-orifice; one 1 cm and the other 3.5 cm in diameter, separated by a fibrotic band (Figs. 3A and 3B). There were 2 underlying papillary muscles. Mitral valve was incompetent and

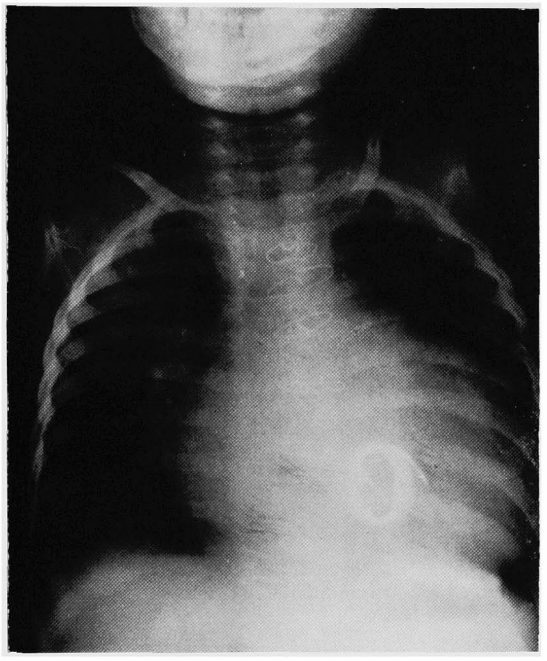


Fig. 4

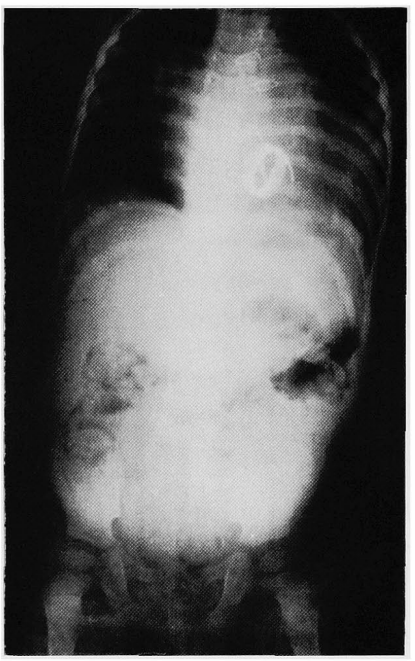


Fig. 5

Fig. 4. Postoperative chest X-ray of the patient with Lillehei-Kaster mitral prosthesis.

Fig. 5. Postoperative X-ray of the child's body. Size of the valve (Lillehei-Kaster prosthesis) and size of the child's body are shown, respectively.

deformed. Both orifices were excised and replaced by No. 20 Lillehei-Kaster mitral prosthesis using continious 2-0 ti-cron sutures. He did well during the postoperative period and kept on dipyridamole (Persantin) 10 mg/Kg and aspirin 50 mg/Kg for the prevention of thromboembolic complications. He was also on a daily dose of digitalis but seldom on diuretics. Control chest X-ray during the postoperative phase displayed a cardiac shadow diminished in size compared to the preoperative X-ray (Fig. 4). Fig. 5 shows an X-ray of the body of the child and the size of the valve, respectively.

He is still under our clinical control and doing well without any complications.

Discussion

Valve replacement in childhood is not common and performed mostly for rheumatic valvulopathies in underdeveloped and developing countries, and for congenital valvulopathies in developed areas of the world.[6),7)] Difficulties in anticoagulation and related problems are the major handicaps after valve replacement in childhood, necessitating the use of anti-platelet

agents (such as Persantin and aspirin) in some of these cases.[6),7)] We have observed in our clinical practice that the need for re-replacement parallel to the child's growth is not a valid point in valve replacement in children, because it is most of the time possible to insert quite a large valve to a child's heart, especially in the mitral position.[7)]

Double-orifice mitral valve with significant regurgitation is one of the indications for valve replacement in children. The fact that the duplication of the mitral valve is associated with partial atrioventricular canal in about 25% of the cases strongly suggests that this malformation is related to endocardial cushion maldevelopment.[4)] Our previous case also supports this theory.[5)]

Duplication of the mitral valve is associated with additional pathologies in 54% of the cases.[4)] Pyorala et al (quoted by Mercer and Tubbs)[1)] listed the associated lesions in order of frequency, as follows: atrioventricular canal, bicuspid aortic valve, coarctation of the aorta, patent ductus arteriosus, right sided aortic arch, subaortic stenosis, bicuspid pulmonary valve, Ebstein's anomaly and secundum atrial septal defect.

The mitral orifices may be either equal or unequal in size.[4)] Combined area of the double-orifices may be smaller than the normal mitral area. Additional number of papillary muscles and chordae tendineae may be present.[4)]

Double-orifice mitral valve can sometimes be overlooked during the operation for atrial septal defect.[3),5)] Cases with primum atrial septal defect should especially be examined for the presence of this malformation.[5)]

No surgical approach had been reported for this congenital lesion until Reed et al's presentation.[3)] This lesion has been reported as an incidental postmortem finding in most of the cases in the literature.[3),8)–10)] Reed et al[3)] divided the bridge of fibrotic tissue between 2 orifices and obtained a satisfactory result. Mercer and Tubbs[5)] presented a case of duplication of the mitral valve associated with subaortic stenosis and closed the smaller orifice by interrupted sutures. Pernot[11)] reported a case of double orifice mitral valve subjected to valve replacement. We did not perform any surgical intervention for the case previously published from our clinic owing to the hemodynamically insignificant mitral regurgitation.[5)] We believe that mitral valve replacement should be the treatment of choice in cases with significant mitral regurgitation. In cases of mitral stenosis, division of the separating band between the orifices should be tried first.[5)]

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