Congenital Mitral Insufficiency with Cleft Posterior Leaflet

By Oscar Creech, Jr., M.D., Marion K. Ledbetter, M.D., and Keith Reemtsma, M.D.

MITRAL INSUFFICIENCY in congenital heart disease most frequently occurs with partial or complete atrioventricular canal defects. In such instances the anatomic lesion consists of a cleft in the anterior leaflet of the mitral valve. Mitral insufficiency as an isolated lesion in congenital heart disease is uncommon. In a recent report Starkey described two cases of isolated congenital mitral insufficiency, and in both instances dilatation of the mitral annulus appeared to be the anatomic basis of the mitral insufficiency.

The present report describes a case of congenital mitral insufficiency due to a cleft in the posterior leaflet of the mitral valve corrected by operation. In addition, this patient showed, by hemodynamic studies, an area of supravalvular stenosis of the aorta.

Case Report

This female infant was first admitted to Charity Hospital at the age of 6 months for repeated respiratory infections. At that time a clinical diagnosis of congenital heart disease was made on the

Figure 1

Left. Anteroposterior roentgenogram of the chest reveals gross cardiac enlargement, particularly of the left atrium and left ventricle. Right. Enlargement of the left atrium is demonstrated on the lateral projection with barium in the esophagus.
Simultaneous measurement of left ventricular and femoral arterial pressures disclosed a systolic gradient of 30 mm. Hg.

Injection into aorta

Injection into left ventricle

Dye-dilution curves were obtained following injection of indocyanine green dye into the aorta with sampling from the femoral artery (top). Subsequent injection into the left ventricle with sampling in the femoral artery (bottom) disclosed curves compatible with mitral insufficiency.

A pullback tracing from left ventricle into aorta disclosed no change in systolic pressure at the aortic valve and demonstrated slight supravalvular obstruction.

step-up of blood oxygen content from the superior vena cava to the right atrium of questionable significance, but there was no evidence of arterialization in the right ventricle or pulmonary artery, Pulmonary wedge pressure was 15/7, with a mean of 13 mm. Hg. A catheter was passed from the brachial artery into the left ventricle, where pressure was recorded at 123/8 with simultaneous femoral artery pressure of 93/68 (fig. 2). A pullback tracing revealed no change in systolic pressure at the aortic valve, but a gradient was found in the supravalvular area (fig. 3). Dye-dilution curves were obtained with injection into the left ventricle and sampling in the femoral artery, and subsequent injection into the aorta with sampling in the femoral artery (fig. 4). These curves were interpreted as suggestive of mitral insufficiency.

On December 19, 1959, the patient was operated upon through a right anterior thoracotomy with

basis of cardiomegaly and a loud systolic precordial murmur. Because of repeated episodes of pneumonia despite digitalization she was readmitted at age 23 months for diagnostic studies. Physical examination revealed a grade III systolic murmur over the entire precordium, loudest at the apex, with radiation to the left axilla. The liver edge was palpated 3 cm. below the costal margin. X-rays revealed gross cardiomegaly, with marked enlargement of the left atrium and left ventricle (fig. 1). Electrocardiograms were interpreted as showing left ventricular hypertrophy.

On November 10, 1959, combined right and left heart catheterization was performed. There was a
The cleft in the posterior leaflet of the mitral valve is shown in drawing (a). The cleft extends from the margin of the leaflet to the annulus. The chordae tendineae to the lesser half of the posterior leaflet were fused and shortened and attached to a large papillary muscle (b). The cleft was closed with interrupted sutures (c and d).

Her postoperative course was smooth, and she was discharged on January 7, 1960. She failed to receive digitalis following discharge from the hospital and was re-admitted on January 19, 1960, in mild failure for redigitalization. Subsequent to this she has been maintained on digitalis. When seen in her eighteenth postoperative month, she was asymptomatic and had normal exercise tolerance. However, on physical examination she had a grade III systolic apical murmur and on x-ray the enlarged left atrium persisted, although left ventricular enlargement appeared less pronounced (fig. 6).

Discussion

Congenital mitral insufficiency most commonly occurs with endocardial cushion defects, such as partial or complete atrioventricular canal. In such cases, there is persistence of the atrioventricular groove and, in addition, failure of fusion of the left tubercles of the anterior and posterior endocardial cushions, resulting in a cleft in the septal leaflet of the mitral valve. In the present case, however, the atrioventricular septum, the interventricular and interatrial septa, and the septal leaflet of the mitral valve all appeared normal. The cleft occurred in the posterior mitral
leaflet, and the mitral valve resembled anatomically the right atrioventricular (tricuspid) valve.

Edwards and Burchell\textsuperscript{2} have described a type of congenital mitral insufficiency resulting from anomalous chordal insertion. In their experience, this lesion occurred most often in association with ventricular septal defect, but it may occur alone. The chordae connecting with the posterior leaflet were usually unduly short, and their lower ends were attached more basally than normal. They were often associated with accessory clefts or commissures in the posterior leaflet. From an examination of these hearts it appeared that the chordae exerted a restraining influence on the leaflet and prevented proper movement for closure of the accessory commissure. Anomalous insertion of chordae may occur also in corrected transposition of the great vessels. In this condition the left atrioventricular valve is a mirror image of the normal tricuspid valve. Mitral insufficiency may result from restricted mobility of the septal and posterior leaflets due to shortened chordae, or from a valvular malformation which is a counterpart of Ebstein’s malformation of otherwise normal hearts.\textsuperscript{2}

In the case reported here it was observed that separate papillary muscles were attached to the two sections of the posterior leaflet by very short chordae. Following closure of the cleft it appeared that complete competency of the valve had been restored, but the patient’s subsequent course, and particularly the persistence of left atrial enlargement and a loud systolic murmur at the apex, suggest that some degree of insufficiency persists. This is due, perhaps, to the shortened chordae to this leaflet. Edwards and Burchell\textsuperscript{2} have de-
scribed an altered relationship between the papillary muscles and valve leaflets when left ventricular enlargement occurs as a result of mitral insufficiency. This tends to augment insufficiency resulting from loss of valve substance. Thus, "mitral insufficiency begets mitral insufficiency." Such a mechanism may be a secondary etiologic factor in this case.

Summary
A case of congenital mitral insufficiency in a 2-year-old infant is reported. No other intracardiac defect was found, although pressure studies suggested an area of supravalvular aortic stenosis.

Diagnostic studies included combined right and left heart catheterization. Dye-dilution curves confirmed the presence of mitral insufficiency.

The anatomic defect, a cleft posterior leaflet of the mitral valve, was repaired by direct suture of the valve leaflet.

During 18 months of postoperative observation, the child has shown symptomatic improvement and decrease in the size of the left ventricle, although a systolic mitral murmur and left atrial enlargement persist. Residual insufficiency is probably due to short chordae that restrict mobility of the posterior leaflet.

References

Text and Footnote
The first time I heard Professor Sarton expound his doctrine privately, he was in his most optimistic mood. I, as a young man, understood him—or perhaps misunderstood him—to say that the future historian of science would write the history of a century solely in cultural terms and largely in terms of the labors of the scientists and scholars. The kings and queens, the politicians, and especially the military campaigns were to be reserved for footnotes, a strong contrast to the usual custom of historians, Sarton pointed out. Orthodox historians might at best insert a footnote to the political history of Great Britain in the eighteenth century to the effect that one Sir Isaac Newton, the Master of the Mint under William III, enunciated his laws of motion and "explained" the workings of the solar system. He, Sarton, proposed to reverse the scheme!—JAMES B. CONANT. History in the Education of Scientists. American Scientist 48: 530, 1960.
Congenital Mitral Insufficiency with Cleft Posterior Leaflet
OSCAR CREECH, JR., MARION K. LEDBETTER and KEITH REEMTSMA

Circulation. 1962;25:390-394
doi: 10.1161/01.CIR.25.2.390

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1962 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/25/2/390