PATENT DUCTUS ARTERIOSUS WITH MITRAL STENOSIS

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An apical mid-diastolic murmur is sometimes audible in patients with a patent ductus arteriosus (Nadas and Alimurung, 1952) but differentiation from organic mitral stenosis is usually not difficult. The patient described in this paper presented with typical murmurs of isolated mitral stenosis and the correct diagnosis of patent ductus arteriosus and trivial mitral stenosis was made only at operation.

Case Report

The patient, a young woman of 19, gave no history of rheumatic fever or chorea. She was first seen by Professor Crichton Bramwell at the age of 6 in 1943. No record is available of the findings at that time except that a standard lead electrocardiogram showed left axis deviation. She was seen again by the same physician in 1949, when the classical physical signs of mitral stenosis were found and the fluoroscopic findings were described as those of an uncomplicated mitral lesion. His comment on the case was "Only the fact that the cardiac abnormality was noticed so early in life made me think there must be an associated congenital lesion."

Between 1949 and 1956 the patient was examined on separate occasions by six different observers all of whom considered she had the classical signs of mitral stenosis. These physical signs were as follows.

There was no cyanosis and normal sinus rhythm. The blood pressure was 110/70–120/80. The cardiac impulse was diffuse and palpable almost out to the anterior axillary line. An apical diastolic and pre-systolic thrill was palpable. On auscultation the mitral first heart sound was loud and there was an opening snap. The pulmonary second sound was loud and split. A loud, full mitral diastolic murmur with pre-systolic accentuation was present and there was a soft apical systolic murmur. On one occasion only, a soft diminuendo diastolic murmur was heard at the lower end of the sternum. The lungs were always clear and there were no signs of congestive heart failure.

Fluoroscopy on several occasions showed the heart to be moderately enlarged and of mitral configuration. Moderate left atrial enlargement was demonstrated and the lung fields showed increased vascularity but no hilar dance.

The electrocardiogram showed slight depression of the S–T segment in aVL and in lead I and although a large R wave was present in V3R there was no other evidence of right ventricular enlargement. Some delay (0.06 sec.) in the onset of the intrinsic deflection, suggesting left ventricular enlargement, was present in V4 and V6 but this was not noted until the record was re-examined after operation.

Towards the end of 1955 the patient began to notice increasing breathlessness on exertion but had no paroxysmal dyspnoea or hæmoptysis. Her incapacity steadily increased until April, 1956, when mitral valvotomy was advised.

At thoracotomy Mr. W. F. Nicholson noted considerable left ventricular enlargement and a grossly dilated pulmonary artery over which a continuous thrill was palpable. The pulmonary veins were large and there was moderate left atrial enlargement. Palpation of the mitral valve revealed a lightly calcified valve, only slightly stenosed and approximately 3 cm. in diameter. The lateral commissure split easily to give a valve of 3.5 cm. No attempt was made to split the medial commissure. Thorough palpation of the left atrium revealed no atrial septal defect; but after further dissection a broad patent ductus arteriosus was isolated and ligated with cessation of the thrill over the pulmonary artery. Seven hours after operation she collapsed due to bleeding which ceased when the ductus was divided and sutured.

All murmurs disappeared immediately after operation and the heart sounds became normal. Six months after operation her exercise tolerance, heart size, and pulmonary vascularity were within normal limits (Fig. 1B).

FIG. 1.—Teleradiograms of patient. (A) Showing cardiac enlargement and increased pulmonary vascular markings extending well out into the lung fields. (B) After ligation of the ductus, showing diminution of heart size and return of pulmonary vascularity to normal. Elevation of the left hemidiaphragm is due to damage to the phrenic nerve at the second operation.

Discussion

Bramwell (1943) pointed out that the production of an obstructive murmur depends on the degree of obstruction relative to the velocity of the blood current: he suggested that an increased rate of blood flow through a normal mitral orifice may be instrumental in producing accentuation and roughening of the first heart sound in thyrotoxicosis and in other conditions in which the heart is over-acting. Patent ductus arteriosus is associated with a large pulmonary flow and an increased flow through the mitral valve, and the apical mid-diastolic murmur not uncommonly heard in the uncomplicated patent ductus probably arises as a result of this (Nadas and Alimurung, 1952). These signs may be regarded as signifying a relative mitral stenosis (Bramwell, 1943) and disappear when the ductus is ligated. In our patient the mitral valve was slightly stenosed and it seems likely that a large increase in blood flow through this slightly narrowed valve caused the auscultatory findings of mitral stenosis.

We cannot explain the absence of the typical ductus murmur. In the presence of pulmonary hypertension the typical murmur may be modified or absent (Whitaker *et al.*, 1955) and the diagnosis difficult or impossible to make without the aid of cardiac catheterization or angiocardiography. Possibly our patient had some degree of pulmonary hypertension as a result of the increased pulmonary blood flow and the mitral lesion, but the evidence suggests that the shunt was predominantly left to right.

Although the ductus was silent three atypical features should have aroused our suspicion: the early age at which well-developed mitral stenosis was found, the signs of left ventricular enlargement in the cardiogram without mitral regurgitation, and the evidence on radioscopy of increased pulmonary blood flow.

The aetiology of the mitral lesion remains unknown. No previous history of rheumatism or chorea was obtained and sections of the atrial appendage removed at operation showed no evidence of rheumatic activity.

Summary

A case of patent ductus arteriosus with slight mitral stenosis presenting with signs simulating severe mitral stenosis has been described. The correct diagnosis was established only at operation. All physical signs disappeared after ligating the ductus and slightly increasing the size of the mitral valve by valvotomy.

Our thanks are due to Professor Crighton Bramwell for allowing us to quote his original report and to Dr. A. Morgan Jones, Mr. W. F. Nicholson, and Dr. E. G. Wade for help and advice in the preparation of this report, and for permission to publish the case.

References

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EDITORIAL NOTE

This case recalls a somewhat parallel example that occurred in the clinic of Professor Gustav Nylin in the Sodersjukhuset, Stockholm, some years ago. It was demonstrated to one of the editors (I.G.W.H.) in 1951, who was greatly impressed at the time by the fact that this error in diagnosis was exhibited and discussed in detail by so eminent a clinician. The case has not been published and we are enabled through the courtesy of Professor Nylin to give the following details.

The patient was a lady of 38 who came under observation in 1947 with a clinical diagnosis of mitral stenosis. There was a long diastolic murmur confirmed by phonocardiogram and she had a blood pressure of 145/98. There was an enormous bulging pulmonary artery on X-ray which was considered due to an atrial septal defect. In 1949 she was admitted after three weeks' illness with low fever, and was regarded as a case of subacute bacterial endocarditis. The apical diastolic murmur was noted to be of a rumbling character. Blood cultures were negative. Her temperature resolved with penicillin but she died suddenly shortly afterwards. The ante-mortem diagnosis was thrombosis or embolus of a pulmonary artery with Lutembacher's syndrome.

At necropsy there was, in fact, thrombosis of the left pulmonary artery and a congenital mitral stenosis. There was no atrial septal defect but a large persistent ductus arteriosus.