A Pathognomonic Murmur of "Atypical" Patent Ductus Arteriosus*

Talma Rosenthal, M.D. and Itzhak Kario, M.D.

A case of patent ductus arteriosus with pulmonary hypertension is presented. The presence of a distinctive rough prolonged diastolic murmur, instead of the usual Graham-Steell murmur of pulmonary incompetence, is stressed. It appears that this finding is typical of a particularly wide and short patent ductus and is classified as typical for atypical patent ductus arteriosus.

The usual case of patent ductus arteriosus is associated with the familiar "machinery-murmur." With the development of pulmonary hypertension and the equilibration of aortic and pulmonary arterial pressures, the machinery murmur is usually replaced by a systolic murmur. If a diastolic murmur is present under these circumstances, it is usually the Graham-Steell murmur, namely a decrescendo, protodiastolic, soft blowing murmur heard best at the pulmonary area and to the left of the sternal margin.

It is not widely appreciated that the development of severe pulmonary hypertension in cases of patent ductus may result in a rough pandiastolic murmur with certain specific features that are quite different from those of the Graham-Steell murmur.

The former is a very harsh holodiastolic murmur, which is not decrescendo in type. Because of its special qualities it may be mistaken for a systolic murmur, even by experienced clinicians.

From the time this murmur was first mentioned in the early 1950's, it several years elapsed before it was established as a sign of severe pulmonary hypertension in cases of patent ductus arteriosus (PDA). However, the English literature does not seem to be aware of this finding, as shown by an extensive search for it.

The following case is presented in order to describe this specific auscultatory finding in greater detail.

CASE REPORT

The patient was a 57-year-old man known to have had congenital heart disease from an early age. Although he had required digitalis for several years, the patient was able to climb stairs without difficulty and to pursue his profession as a watchmaker until two months before admission.

Physical examination revealed a thin adult man with mild peripheral and central cyanosis. There was no clubbing of digits. He was markedly dyspneic, but lying flat in bed did not distress him any further. The neck veins were markedly distended, and extended up to the angle of the jaw. Clearly visible jugular "a" waves and prominent carotid pulsations were noted. The pulse was regular and collapsing. The blood pressure was 140-150/60. There was mild pitting edema of the legs without any difference in color between the upper and lower limbs. The apex impulse was left ventricular in type, and was palpated in the 7th intercostal space, at the anterior axillary line. There was a marked right ventricular lift. A very easily palpable diastolic thrill was present at the apex and was also felt at the left of the sternal margin. The first sound was slightly accentuated at the apex, and a short grade II systolic murmur was present. A booming rough diastolic murmur particularly grating to the ear was the distinctive auscultatory finding. The murmur was pandiastolic (starting immediately after the second sound and continuing until the next first heart sound). It was best heard at the lower end of the sternum. The second heart sound was closely split and not accentuated. There were mild crepitations at both lung bases, and a tender liver was palpable 8-10 cm below the right costal margin. Phonocardiogram confirmed the auscultatory findings (Fig 1). The electrocardiogram showed sinus rhythm and a rate of 80 per minute. There was right axis deviation (+120°). The P waves in the limb leads were diphasic. High sharp P waves and an rsR' pattern were present in the right ventricular leads. There were deep S waves in V1-V5 with the transitional zone at V6. The electrocardiogram was thought to be consistent with biventricular hypertrophy. Chest roentgenograms and fluoroscopy showed hilar dance with dilated proximal vessels

*From the Cardiological Institute and Tel-Aviv University Medical School, Tel-Hashomer Government Hospital, Tel-Aviv, Israel.

Figure 1. Pandiastolic hand-like murmur overlapping the first heart sound.
DISCUSSION

The diastolic murmur of our patient was unusually rough and vibrant and was accompanied by a thrill. It was heard immediately after the second heart sound and continued throughout diastole, obliterating the subsequent first heart sound. The murmur was so rough that even experienced clinicians mistook it for a systolic murmur, until the phonocardiogram proved otherwise. This type of murmur was first mentioned in 1950 by Johnson, but was not fully characterized until Holldack's description in 1957. In 1962, Fishleder et al reviewed the cases described in the literature and added five of their own. In all these patients the important findings consisted of a rough diastolic murmur accompanied by a thrill and associated with pulmonary hypertension and evidence of a bidirectional shunt.

In all hemodynamic studies, the pressure in the pulmonary artery approached that of the aorta.

In every instance, post mortem examination revealed an unusually short and wide patent ductus arteriosus, with signs of pulmonary hypertension.

![Figure 2](upper) and 3 (lower). Heart is enlarged with marked prominence of the pulmonary conus and some dilatation of the aorta. The left ventricle is enlarged. Note the paucity of the peripheral vasculature.

and poor peripheral vascular markings. There was a marked increase in the transverse cardiac diameter in the postero-anterior view. The left oblique view revealed a predominantly enlarged left ventricle. A clearly defined prominent pulmonary arc and mild unfolding of the aorta were seen. Tomography showed calcification of the aortic knob as well as of the main pulmonary trunk and left pulmonary artery (Fig 2 and 3). The tentative diagnosis was "patent ductus arteriosus," and the patient was admitted for treatment of his cardiac failure and further investigation by cardiac catheterization. However, his condition deteriorated and he remained unresponsive to treatment. Unfortunately he died a month after admission. Cardiac catheterization could not be performed because of his poor condition.

At postmortem examination, the heart weighed 730 grams. There was marked hypertrophy and dilatation of both ventricles. The pulmonary artery was tremendously dilated and atheromatous, and the pulmonary ring measured 10 cm in diameter. An unusually short and wide patent ductus (2 cm in diameter) was present (Fig 4 and 5). Two cm below the pulmonary valve there was an area of thickened endocardium, which included four regurgitation pouches (Fig 6).

![Figure 4](left ventricle and aorta, arrow points to patent ductus. Above the defect the aorta is normal, while below, it is widely dilated.)
The cause of the murmur is not clear. Fishleder et al suggested that it is produced by pulmonary insufficiency with backflow of blood to the right ventricle, resulting in a reduction of pressure at the pulmonary ostium. The latter may cause blood to flow from the aorta to the right ventricle, thereby producing a diastolic murmur. According to this concept, the murmur is made up of two components, the first caused by flow from the aorta to the pulmonary artery, and the second being the murmur of pulmonary incompetence.

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Reprint requests: Dr. Rosenthal, Heller Institute, Tel-Hashomer Government Hospital, Tel-Aviv, Israel

Reversed Reciprocating Paroxysmal Tachycardia*

M. C. Mehta, M.D. and S. V. Singh, M.D.

A case report of reversed reciprocating tachycardia sensitive to vagal stimulation which produced interesting patterns of coupling is described. The mechanism of reversed reciprocal rhythm is discussed. This arrhythmia and Wolff-Parkinson-White syndrome can be explained on the basis of anomalous pathway. In the former partial anterograde block through the A-V node is also present.

Katz and Pick have described a cardiac arrhythmia, a form of reciprocal rhythm, with the pace-maker of sinus or auricular origin. Under such a situation the auricular impulse travels to the A-V node and from there to both ventricles and atria producing a P-QRS′ pattern. P′ is the result of

*From the Department of Medicine and Cardiology, R.N.T. Medical College, Udaipur-Rajasthan, India.