Patent Ductus Arteriosus and Pulmonary Valve Insufficiency; Unusual Clinical Manifestations*

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Patent ductus arteriosus is a congenital cardiac lesion which almost always manifests itself as a "machinery" murmur. A small percentage of patients, however, have atypical murmurs. These are usually associated with the presence of other congenital anomalies or the presence of pulmonary hypertension which alters the systolic-diastolic pressure gradient across the defect. The following case demonstrates an unusual murmur in a patient with patent ductus arteriosus and a left-to-right shunt.

CASE REPORT

The patient, a 55-year-old white man who is mute, was admitted to Community Hospital of Indianapolis on November 23, 1968. Much of the history was provided by the patient's mother. The informant was first told of the presence of a heart murmur in her son sometime during his early teens. The murmur was attributed to rheumatic fever, despite no recognizable clinical episode of this illness. His early years were characterized by recurrent episodes of bronchitis and mild effort dyspnea. He had no other significant illnesses. Four months prior to hospitalization the dyspnea increased in intensity and finally the patient was unable to carry on his normal sedentary occupation because of cough and shortness of breath. He had two-pillow orthopnea, paroxysmal nocturnal dyspnea, but no pedal edema or chest pain. In spite of digitalization and intermittent diuretic therapy, these symptoms persisted. Family history failed to reveal other members who were mute or who had any heart disease.

Physical examination upon admission revealed a well-developed, well-nourished man who had a dry, hacking cough and appeared slightly short of breath. The blood pressure was 130/60 in both arms recumbent and the heart rate was 80 and regular. The pulse was bounding but not considered to be "waterhammer" in quality. No definite peripheral findings of a wide pressure were noted. There was no clubbing or cyanosis. A prominent bulge of the left anterior chest from the second to fifth ribs was quite evident. The remainder of the physical examination was not remarkable except for that of the heart. The point of maximal impulse was in the fifth intercostal space at the anterior axillary line. A systolic lift was present along the left sternal border. A diastolic thrill was noted over the second to fifth intercostal spaces to the left of the sternum. The heart sounds were normal except that only one component of S2 could be definitely heard. A grade IV/V holodiastolic murmur was present over the entire precordium. This murmur was maximal, however, in the area of thrill. The phonocardiogram obtained is demonstrated in Figure 1. The quality of this murmur could

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best be described as similar to the murmur of a ventricular septal defect, but of course the timing in the heart cycle was diastolic rather than systolic. Clinically, no other murmurs were heard. Significant laboratory studies were a non-reactive VDRL, hemoglobin of 16.0 gm percent, hematocrit 50, white blood cell count normal, BUN 24 mg percent, two-hour post-prandial blood sugar 120 mg percent, CO₂ content 30 mEq/L, chloride 95 mEq/L, sodium 143 mEq/L, potassium 4.6 mEq/L. Blood cultures on three separate occasions were negative. The cardiac catheterization findings are summarized in Table 1. Oxygen step-up at the pulmonary artery level localizes the left-to-right shunt to this area. Peripheral desaturation was not noted, indicating right-to-left shunting was not present.

Electrocardiogram demonstrated right axis and an incomplete right bundle branch block pattern. No other abnormality was noted. Chest roentgenogram showed evidence of left and right ventricular hypertrophy.

**DISCUSSION**

Persistent patency of the ductus arteriosus is characterized by a left-to-right shunt through the duct and a rather classic "machinery" murmur. In Wood's series of 115 cases, the usual findings were present in 95 percent of the cases. A Gibson murmur is one heard best in the second left interspace, is more or less continuous, waxes toward the end of systole and wanes in mid-diastole. The murmur is not typical when the pulmonary vascular resistance becomes elevated, resulting in a decrease or reversal of the shunt. In these cases the diastolic component of the murmur diminishes and only a faint systolic ejection murmur may remain.

The present case demonstrates an extremely unusual pathophysiologic situation in patent ductus arteriosus which resulted in bizarre physical findings. During systole, there was essentially no pressure difference recorded simultaneously between the aorta and pulmonary artery. A large pulse pressure due to pulmonary insufficiency was present in the pulmonary artery and the diastolic pressure here was significantly lower than that in the aorta (Fig 2). This gradient permitted a left-to-right shunt across the ductus only during diastole.

Phonocardiograms obtained from the right ventricle and pulmonary artery during the cardiac catheterization demonstrated maximal intensity of the murmur in the pulmonary artery at the level of the patent ductus. A faint murmur during diastole was heard in the right ventricle, however. This would indicate that the loud murmur heard on the chest wall was produced by the patent ductus rather than by the pulmonary insufficiency.

Right atrial cineangiogram demonstrated an extremely large main pulmonary artery. Injection of cardiogreen dye into the pulmonary artery and sampling of the dye in the right ventricle revealed immediate appearance of the dye consistent with pulmonary regurgitation.

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**Table 1—Cardiac Catheterization Data**

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<th>Pressure mm Hg</th>
<th>% O₂ Saturation</th>
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<tr>
<td>IVC</td>
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<tr>
<td>RA</td>
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<tr>
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<tr>
<td>LV</td>
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<tr>
<td>Aorta</td>
<td>136/70 Mean 90</td>
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</table>

**Figure 2.** Simultaneous pressure tracings recorded in the pulmonary artery and aorta at the level of the ductus arteriosus. Note the diastolic pressure gradient as a result of the wide pulmonary artery pulse pressure, secondary to pulmonary insufficiency.
Atypical clinical findings are seen in 5 percent of patients with patent ductus arteriosus. The size of the ductus, as well as the pressure relationships between the aorta and pulmonary artery, determines the type of murmur present vis-a-vis the existing systolic-diastolic pressure gradients. The diastolic murmur may simulate that of aortic insufficiency. Indeed, Gaugahan et al reported four patients observed for a long time with a presumed diagnosis of aortic insufficiency who eventually were found to have patent ductus arteriosus, pulmonary artery dilatation and resultant insufficiency. However, in these cases the murmurs were distinctly decrescendo blowing murmurs and thus different from the case presented here. Calne and Raftery reported an elderly man with patent ductus arteriosus and a prominent diastolic murmur described as a "long early diastolic murmur which varied with respiration." Pulmonary insufficiency was found to be present, but again the characteristic of the murmur was evidently different from this patient's. In reviewing the literature, four cases similar to this have been described. These patients had patent ductus arteriosus with large pulmonary arteries and pulmonary insufficiency. Harris and Peters' patient was most nearly like ours. Their patient was observed for 18 years. The typical Gibson murmur gradually was replaced by an intense holodiastolic murmur, loudest in the pulmonic area but audible over the entire precordium. The murmur was coarse in character and low-pitched. A diastolic thrill was present and no systolic murmur was heard. Autopsy at age 43 proved the presence of patent ductus arteriosus and pulmonary insufficiency.

SUMMARY

A patient with patent ductus arteriosus is described in whom the combination of pulmonary hypertension and pulmonary insufficiency seem to allow shunting of blood only in diastole. These circumstances produced a peculiar, low-pitched holodiastolic murmur, maximally heard in the second to fourth intercostal space to the left of the sternum. The murmur appeared to be produced by eddy currents in the region of the ductus rather than in the area of the pulmonary valve. The case is reported because of the rarity with which this type of murmur is heard with patent ductus arteriosus.

ADDENDUM: Since the submission of this article, Rosenthal and Kariv have reported a strikingly similar murmur with patent ductus arteriosus. Hemodynamic data were not obtained in their study.

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THE ORIGIN OF HOSPITALS

In Arabic the word for hospital is Maristan. The earliest Maristan in Muslim times is believed to have been built by Al-Walid in the years 705-715. Doctors were assigned to staff the hospital and received salaries. Arabic medicine spread rapidly to other parts of the medieval world, and had a profound influence on the development of similar European institutions. In Europe the hospital was, in fact, a direct outgrowth of the Arabic Maristan. Even modern hospital administration and procedures are largely an outgrowth of that used by the Arabs. The outpatient and inpatient idea, separate wards for some diseases, the method of examination of patients, the utilization of the hospital patients for teaching medical students, regular visits of the chief physicians, pharmacies—all of these came directly to us from the medieval Arabic hospitals.


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