Isolated Pulmonic Valvular Regurgitation

By Ralph F. Morton, M.D., and Thomas N. Stern, M.D.

An extremely rare case of isolated organic insufficiency of the pulmonic valve is reported along with clinical and hemodynamic data.

Isolated nonsurgical pulmonic valvular regurgitation has been reported only twice during life in man. In fact, it is unusual to make a diagnosis of organic pulmonic insufficiency from any cause with any degree of certainty during life. The following case is presented as one of symptomatic heart disease in an adult secondary to pulmonic valvular regurgitation.

Case Report

V. H., a 20-year-old Negro housewife, was apparently normal at birth except for small super-numerary digits on each of her 4 extremities. As a child she was able to keep up with other children until her early teens when she noted easy fatigability on moderate exertion and inability to run because of dyspnea and marked dizziness. At 15 years of age she had pleurisy and shortness of breath for about 2 months. When seen by a physician, she was told that she had “heart trouble probably from birth.” Shortly thereafter she had severe tonsillitis, but no rheumatism, chorea, or other stigmata of rheumatic fever. Her first pregnancy at 15 years of age terminated spontaneously at 3 months. During her second pregnancy at 20, moderate exertional dyspnea was noted, but delivery of a normal male infant was tolerated without evidence of heart failure. The patient had her first complete physical examination 3 months after her second pregnancy, when she developed cough, fever, and pleurisy of the right chest. In addition to evidence of pneumonitis, she was noted to have a heart murmur. After the pneumonitis responded to antibiotics, the patient was referred to the Cardiovascular Clinic of the City of Memphis Hospitals because of her murmur, mild dyspnea on exertion, and dizziness on moderate exertion.

The patient was normally developed. There was no dyspnea, cyanosis, or clubbing of the fingers. The pulse was 72 and the blood pressure 110/70. There was slight decrease in breath sounds over the right lower chest, but the chest and lungs were otherwise normal. The cardiac apex was in the fourth inter-costal space, just medial to the midclavicular line. A moderate systolic impulse was noted along the left sternal border, and a diastolic thrill was localized at the pulmonic area. The pulmonic and apical second sounds were split. A soft, grade 2, moderately high pitched, holosystolic murmur was heard at the base of the heart, but was maximal at the pulmonic area. An early, rough, loud, decrescendo diastolic murmur was heard best at the second and third left intercostal spaces but also radiated along the left sternal border and to the midprecordium. The peripheral pulses were normal.

An electrocardiogram (fig. 1) revealed right bundle-branch block. The phonocardiogram (fig. 2) confirmed the murmurs previously described. The chest x-ray showed no increase in heart size, but prominence of the pulmonary artery was noted. Marked pulsations of the right ventricle and primary branches of the pulmonary artery were demonstrated on fluoroscopy and roentgenkymography (fig. 3). Pulsations of the pulmonary artery were much more prominent than those of the aorta.

Diagnoses considered prior to cardiac catheterization were atrial or ventricular septal defect with pulmonic insufficiency and patent ductus arteriosus.

Cardiac catheterization was performed on February 1, 1956. The catheter was introduced through a left antecubital vein, took the normal course through the pulmonary artery, and was wedged without difficulty. Marked whining of the catheter was noted in the main pulmonary artery. Catheterization findings are summarized in table 1.

Right atrial mean pressure was within normal limits. Right ventricular and pulmonary artery systolic pressures were normal and essentially the same. The pulmonary arterial diastolic pressure was low and not significantly different from the right ventricular end-diastolic pressure. There was an abrupt steep slope of the catabolic limb of the pulmonary pressure curve (fig. 4). This tracing was reproducible and, we believe, not an artifact. Oxygen saturations, oxygen consumption, cardiac output, cardiac index, pulmonary arterial flow, and pulmonary and peripheral resistances were all normal at rest.
ISOLATED PULMONIC VALVULAR REGURGITATION

Fig. 1. Electrocardiogram showing right bundle-branch block.

Fig. 2. Phonocardiogram recorded at pulmonic area showing a soft, high pitched, holosystolic murmur and a loud, coarse, early diastolic murmur.

**TABLE 1.—Catheterization Data**

<table>
<thead>
<tr>
<th>Location</th>
<th>Oxygen content (vol. %)*</th>
<th>Pressures (mm. Hg)†</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior vena cava</td>
<td>9.9</td>
<td>—</td>
</tr>
<tr>
<td>Right atrium</td>
<td>9.9</td>
<td>Mean 3.2</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>10.1</td>
<td>22/3</td>
</tr>
<tr>
<td>Main pulmonary artery</td>
<td>9.9</td>
<td>21/2 Mean 8.9</td>
</tr>
<tr>
<td>Right pulmonary artery (distal)</td>
<td>10.3</td>
<td>17/2 Mean 7</td>
</tr>
<tr>
<td>Pulmonary wedge</td>
<td>—</td>
<td>Mean 4.2</td>
</tr>
<tr>
<td>Brachial artery</td>
<td>14.0</td>
<td>104/68 Mean 80</td>
</tr>
</tbody>
</table>

* Determined according to the technic of Van Slyke and Neill.
† Recorded with a Statham strain-gage manometer and Sanborn recorder.
‡ Measured with a Collins Respirometer.

Fig. 3. Roentgenkymogram showing marked pulsations of the main pulmonary artery that are greater than those of the aorta.
DISCUSSION

Whereas isolated valvular pulmonic stenosis has in recent years been recognized to be a relatively common congenital anomaly, other organic pulmonary valve lesions, either congenital or acquired, are very rare. Organic pulmonary insufficiency due to congenital absence of valve cusps and due to an abnormal number of cusps, 2, 4, or 5, has been reported from autopsy material. Recently, pulmonary insufficiency was conclusively diagnosed during life in a patient with pulmonic stenosis and an interventricular septal defect. Rare causes for acquired pulmonic regurgitation are rheumatic fever, syphilis, acute bacterial endocarditis, and trauma to the chest.

Functional pulmonic insufficiency due to any condition resulting in dilatation of the pulmonary artery with stretching of the valve ring is generally considered to be quite common; however, statistics regarding its frequency are varied and unreliable. This is, of course, never an isolated abnormality.

The diagnosis of pulmonic regurgitation has in the past been based on the presence of a soft blowing diastolic murmur heard best in the second or third left intercostal space, and has required the absence of peripheral signs of aortic regurgitation. It is now generally recognized that this in itself is not adequate evidence for a diagnosis of pulmonic insufficiency. At the present time one is no longer handicapped by the lack of objective methods for making a diagnosis of pulmonic insufficiency during life. Fluoroscopy, by demonstrating a collapsing pulse or hilar dance of the pulmonary vessels, is a valuable aid. An electrokymographic tracing of the pulmonary knob reveals high pulsations that are larger than those of the aorta. In addition, the pulsations have a rapid rise and collapse. Cardiac catheterization probably provides the best evidence for the presence of pulmonic regurgitation. A widened pulmonary artery pulse pressure may be revealed. However, as pointed out by Kohout and Katz, "More important, in that it establishes the diagnosis of pulmonic insufficiency, is the abrupt slope of the cataractism to a value practically identical with that in the right ventricle, and the remainder of the diastolic portion of the pulmonary artery pressure curve is horizontal."

Very few instances of pulmonic insufficiency have been conclusively diagnosed during life. Pulmonic insufficiency associated with other cardiac abnormalities has been diagnosed by means of cardiac catheterization; however, to our knowledge, only 2 cases of isolated nonsurgical pulmonic valvular regurgitation diagnosed ante mortem have been reported.

In our case, the murmur and its phonocardiographic picture, the fluoroscopic findings, and the pulmonary artery pressure curve are quite compatible with a diagnosis of pulmonary valvular insufficiency without associated defect. It is of interest to note that the electrocardiogram showed evidence of right bundle-branch block. Though this pattern is not specific for pulmonic regurgitation, it is the expected finding according to the concept of Cabrera that diastolic overloading of the right ventricle causes right bundle-branch block. It cannot be stated with certainty whether this is a congenital or an acquired defect of the valve, since the patient was not examined prior to 15 years of age.

There is no guide to the prognosis of this apparently pure pulmonic valve lesion, inas-
much as the only other reported cases were those of a 7-year-old child who was asymptomatic\(^1\) and a 44-year-old woman who developed congestive heart failure and died of unknown cause.\(^2\) The latter case, however, was complicated by the presence of diffuse pulmonary fibrosis secondary to tuberculosis. We believe our case to be the only reported instance of uncomplicated, isolated pulmonic valvular regurgitation with symptoms. Even experimentally produced pulmonic regurgitation\(^3\) and that resulting from surgical correction of pulmonic stenosis\(^4\) after short-term follow-up failed to produce right heart failure. We will, therefore, watch our patient’s subsequent course with great interest.

**Summary**

A case of isolated nonsurgical pulmonic regurgitation, believed to be the third instance diagnosed during life, is presented. Diagnosis was suggested by physical examination, phonocardiography, and fluoroscopy and was definitely established by cardiac catheterization.

**Summario in Interlingua**

Es presentate un caso de isolate non-chirurgic regurgitation pulmonic. Secundo le informaciones del autores, isto es le tertie tal caso con establimento del diagnose durante le vita del paciente. Le diagnose esseva suggerite per le examine physic e per phonocardiographia e fluoroscopia. Illo esseva definitivemente establite per catheterisate cardiae.

**References**

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