Isolated Incompetence of the Pulmonic Valve

By Ben O. Price, M.D.

IN 1936 Kissin reviewed the literature and described the clinical and pathologic picture of isolated incompetence of the pulmonic valve. His study was based upon an analysis of autopsy protocols, and it remained for Kezdi et al., in 1955, to report the first case diagnosed during life. To date 13 cases of isolated incompetence of the pulmonic valve have been reported in the English literature.

It is the purpose of this paper to report two additional patients diagnosed during life, and to summarize the data available on these 15 cases.

Case Reports

Case 1

A. A., a 60-year-old woman, was first told that she had a heart murmur at the age of 15. During the next decade she experienced two uncomplicated deliveries. At the age of 27 she was refused life insurance because of heart disease. Her first cardiac symptoms developed at the age of 50, when she had an episode of rapid heart action, shortness of breath, and anxiety. In May 1957 she had moderately severe dyspnea on exertion. Other symptoms strongly suggested that emotional tension might be the major factor in production of the dyspnea.

Examination at that time revealed a blood pressure of 138/90. Abnormal physical findings were limited to the heart. The rhythm was regular and there were no thrills or abnormal pulsations. A grade-II systolic murmur and a grade-I harsh, diastolic murmur were best heard along the left sternal border in the third interspace. The first heart sound was split in the mitral area. An electrocardiogram was interpreted as right ventricular hypertrophy. Fluoroscopy demonstrated enlargement of the left atrium and very active pulsation of the hilar vessels.

At right heart catheterization on May 8, 1957, the pulmonary artery and right ventricular pressures were identical, and there was no evidence of a left-to-right shunt (table 1).

She was seen at the University of Arkansas Medical Center in May 1959, complaining of numbness and paresthesia of the arms and hands, insomnia, and shortness of breath on exertion. She stated that she had been taking digitalis for the past 2 years and had been able to work steadily, although occasionally she felt weak and dyspneic. Except for two episodes of pneumonia there was no history of infectious disease, rheumatic fever, or syphilis.

Positive physical findings again were limited to the heart. There were no palpable thrills and the rhythm was regular. A grade-II systolic murmur was best heard in the second left intercostal space parasternally, and a grade-III rough, crescendo-decrescendo, middiastolic murmur was heard in the same area. Routine laboratory studies were normal.

An electrocardiogram (fig. 1) showed sinus rhythm and right ventricular hypertrophy. Chest fluoroscopy confirmed the presence of active hilar pulsations and generalized cardiac enlargement, predominantly involving the right ventricle and pulmonary artery.

Right heart catheterization was repeated on August 26, 1959 (table 1).

Case 2

E. M., a 37-year-old Negro woman, was referred to the hospital for evaluation of hypertension. She complained of dyspnea on exertion and palpitation. At the age of 15, 3 months after the delivery of her first child, she developed dyspnea, orthopnea, and anasarca. She was told that she had a large heart and was given digitalis. After 8 months of convalescence she was asymptomatic and medications were discontinued.

During the next 10 years she had four more pregnancies, the first two being uncomplicated. Her fourth pregnancy was complicated by the appearance of hypertension. She remained asymptomatic, however, until the last trimester of her fifth pregnancy, when she developed shortness of breath with an inordinate amount of ankle edema. The labor, delivery, and postpartum period were uncomplicated. For the next 5 years she had minimal cardiac symptoms and was able to perform her housework with little limitation of activity. The antecedent history revealed that the patient’s mother and father had died of hypertension.

Examination showed a moderately obese, alert Negro woman whose blood pressure ranged between 120/30 and 150/100 mm. mercury. Abnormal physical findings were limited to the heart, which was moderately enlarged to the left. There were no thrills and the rhythm was regular. A

From the Department of Medicine, University of Arkansas Medical Center, Little Rock, Arkansas.

Present address: U. S. Naval Hospital, Philadelphia, Pennsylvania.

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INCOMPETENCE OF THE PULMONIC VALVE

597

Data on Two Patients

<table>
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<tr>
<th>Position</th>
<th>RV</th>
<th>PA</th>
<th>BA</th>
<th>IVC</th>
<th>RA</th>
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<td>45/5</td>
<td>45/5</td>
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<td>12/2</td>
<td>10/0</td>
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<td>40/4</td>
<td>156/84</td>
<td>2†</td>
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<td>62</td>
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<tr>
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<td>—</td>
<td>124/85</td>
<td>3*</td>
<td>3*</td>
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<tr>
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<td>43/0</td>
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<tr>
<td>Per cent saturation</td>
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</table>

*Millimeters of mercury.
†Mean pressure.
RV, right ventricle; PA, pulmonary artery; BA, brachial artery; IVC, inferior vena cava; RA, right atrium; SVC, superior vena cava.

grade-II systolic and a grade-II harsh, diastolic murmur were heard best in the third left intercostal space parasternally. A phonocardiogram demonstrated a rough, holosystolic murmur loudest over the pulmonic area, a distinct second sound at the pulmonic area, and a crescendo-decrescendo middiastolic murmur of low frequency and moderate intensity.

An electrocardiogram showed right ventricular hypertrophy (fig. 2). Chest fluoroscopy demonstrated a pulsating right hilar mass and moderate cardiac enlargement.

At right heart catheterization in June 1957 (table 1), the pulmonary artery was not entered for technical reasons. In April 1960, repeat right heart catheterization and angiocardiography were performed. The angiocardiogram demonstrated aneurysmal dilatation of the pulmonary arteries. Catheterization (table 1) demonstrated identical pressures in the pulmonary artery and right ventricle with mild systolic hypertension. The pulmonary wedge pressure was normal.

Comment

In both patients heart disease was diagnosed early in life, suggesting the presence of a congenital defect. While both patients have a limited exercise tolerance, neither has objective evidence of heart failure.

Review of Literature

The data of 15 cases diagnosed during life are summarized in table 2. Their ages ranged from 7 to 60 years, with a median of 24 years, and there were eight females and seven males. While nine patients had definite cardiac symptoms, six had complicating diseases that probably contributed to their symptoms. All patients free of cardiac symptoms were under 25 years of age; however, congestive failure did occur in patients who had other diseases.

In every case systolic and diastolic murmurs were heard over the second and third left intercostal spaces at the sternal border. In five cases a diastolic thrill was palpable. Phonocardiograms were described in six cases and all demonstrated murmurs of maximum intensity in middiastole. Five of these were definitely crescendo-decrescendo in character.

Electrocardiograms were reported as normal in four cases. Right ventricular hypertrophy or incomplete right bundle-branch block was diagnosed in seven cases. Low voltage and right axis deviation were each observed once.

Increased pulsation of the pulmonary vessels was the most outstanding roentgenographic finding, being present in every instance in which cardiac fluoroscopy was performed. Enlargement of the pulmonary artery and right ventricle was noted 10 and six times respectively.
### Table 2

**Isolated Incompetence of Pulmonic Valve—Cases Diagnosed During Life**

<table>
<thead>
<tr>
<th>Author and reference</th>
<th>X-ray</th>
<th>Electrocardiogram</th>
<th>Catheterization data</th>
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<tbody>
<tr>
<td></td>
<td>Age</td>
<td>Sex</td>
<td>Over-all heart size</td>
</tr>
<tr>
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<td>24</td>
<td>M</td>
<td>N</td>
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<tr>
<td>Olesen¹, 1955</td>
<td>45</td>
<td>F</td>
<td>Lge</td>
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<tr>
<td>Ehrenhaft³, 1955</td>
<td>14</td>
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<tr>
<td>Ford⁴, 1956</td>
<td>43</td>
<td>F</td>
<td>N</td>
</tr>
<tr>
<td>Morton⁵, 1956</td>
<td>20</td>
<td>F</td>
<td>N</td>
</tr>
<tr>
<td>Segel⁶, 1957</td>
<td>7</td>
<td>M</td>
<td>N</td>
</tr>
<tr>
<td>Dickens⁷, 1958</td>
<td>46</td>
<td>F</td>
<td>Lge</td>
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<tr>
<td>Lendrum⁸, 1958</td>
<td>16</td>
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<td>N</td>
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<tr>
<td>Fish⁹, 1959</td>
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<td>N</td>
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<td>9</td>
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<td>Kjellberg¹¹, 1955</td>
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<tr>
<td>Collins¹², 1960</td>
<td>36</td>
<td>F</td>
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<tr>
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<tr>
<td>Total</td>
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</tr>
</tbody>
</table>

* Diagnosis and autopsy findings:

- **Pulmonic incompetence, congenital**
- **Gonorrheal pulmonic valvulitis**
- **Absence PV—(†) congenital**
- **Bicuspid PV, congenital, pul. fibrosis, myocardial fibrosis**
- **Pulmonic valve incompetence, (†) congenital**
- **Pulmonic valve incompetence, congenital**
- **Bicuspid pulmonic valve, pulmonar arteriolosclerosis**
- **Pulmonic valve incompetence, congenital**
- **Pulmonic valve incompetence, cause unknown**
- **Pulmonic valve incompetence, congenital**
- **Pulmonic valve incompetence, congenital**
- **Withdrawal: dilatation of pulmonary artery and incompetence of pulmonary valve**

- Phonocardiogram; +, yes; (—), no; 0, unknown; N, normal; Lge, enlarged; RVH, right ventricular hypertrophy; CI, cardiac index; PVR, peripheral vascular resistance in dynes per cm.²; RBBB, right bundle-branch block; PA, pulmonary artery; PV, pulmonary valve; RV, right ventricle.
Right heart catheterization was performed in all cases. Nearly identical diastolic pressures in the right ventricle and pulmonary artery were observed in every instance. Several authors emphasized the diagnostic significance of a steeply sloping dicrotic limb in the pulmonary artery pressure curve. When cardiac output was determined, the values were moderately low.

At the time of report, 13 patients were alive and two were dead. Autopsy of both these patients demonstrated congenital deformities of the pulmonic valve that were adequate to produce the insufficiency observed clinically. While diagnoses in the other cases are presumptive, the early ages when heart disease was discovered suggests a congenital etiology.

Discussion

In 1936 Abbott demonstrated that isolated incompetence of the pulmonic valve comprised 0.8 per cent of 1,000 consecutive cases of congenital heart disease. The data presented here demonstrate that this lesion, while rare, has a distinct clinical picture. The diagnosis should be considered in patients with a low-pitched, middiastolic murmur over the base of the heart who have evidence of right heart enlargement. Cardiac fluoroscopy is helpful and may demonstrate enlarged, vigorously pulsatile pulmonary arteries and right ventricular enlargement. The electrocardiogram
Electrocardiogram (case 2) showing right ventricular hypertrophy.

is not distinctive and may be normal. Frequently right bundle-branch block or right ventricular hypertrophy is seen.

The clinical impression can be confirmed by right heart catheterization. The diastolic pressure in the pulmonary artery should be essentially the same as in the right ventricle. A late or absent dicrotic notch and a steeply sloping dicrotic limb are characteristic features of the pulmonary artery pressure curve.

Numerous authors have been concerned with the hemodynamic effects of pulmonic insufficiency. Working with dogs in whom this lesion was produced surgically, Barger, Roe, and Richardson concluded, "pulmonic insufficiency has remarkable little effect on work tolerance or atrial pressure even after months of daily strenuous exercise." Fowler and Dushane demonstrated right ventricular dilatation and a fall in resting cardiac output in eight dogs that survived 11 to 18 months after surgical excision of the pulmonary valve. They concluded that pulmonic insufficiency was not an entirely benign lesion. Blount, McCard et al., reported five patients who developed pulmonic insufficiency after corrective surgery for pulmonic valvular stenosis. In all cases the heart size increased postoperatively. While no hemodynamic alterations were observed, the follow-up time was relatively short.

The auscultatory findings in isolated pulmonic insufficiency are of interest. A systolic
and a diastolic murmur were invariably heard along the upper left sternal border. The second sound was usually of normal intensity and occasionally was split. Phonocardiograms show an interval between the second sound and the onset of the murmur which is of relatively low frequency and has a crescendo-decrescendo quality. The cause of the mid-diastolic murmur is not known. Segel et al., stated that it was "similar in mechanism to the apical middiastolic murmur associated with aortic regurgitation." Since the maximum intensity of the murmur occurs when the right ventricular pressure is lowest, one may speculate that the murmur is produced by rapid filling of the dilated right ventricle.

Our data and that collected from the literature clearly indicate that isolated pulmonic insufficiency is not an entirely benign disease in the human being. The presence of symptoms, evidence of right ventricular enlargement, and low resting cardiac output confirm that isolated incompetence of the pulmonic valve does interfere with normal cardiac function.

As one might predict, any pathologic process that tends to increase pulmonary blood flow or pulmonary artery pressure will further compromise right ventricular function. This was beautifully demonstrated by the case of Smith et al., in which heart failure occurred in a fetus who had no pulmonic valve. The case reported by Ford et al., of pulmonary hypertension developing in a 43-year-old woman affords an example of right ventricular failure occurring with a complicating disease. The dilatation and hypertrophy of the right ventricle and pulmonary outflow tract are best explained by the increase in stroke volume produced by incompetence of the pulmonic valve. These changes may be expected to decrease cardiac reserve to the point that failure of the right ventricle will occur when it is subjected to increased work loads.

**Summary**

Two cases of isolated incompetence of the pulmonic valve diagnosed during life are reported and a review of the literature is presented.

The diagnosis should be suspected in patients with a basal diastolic murmur and right ventricular enlargement who have no evidence of other cardiac valvular disease.

The characteristic murmur associated with isolated incompetence of the pulmonic valve appears to be related to filling of the dilated right ventricle.

The diagnosis can be confirmed clinically by right heart catheterization.

The lesion is not benign in the human being, and right ventricular failure may be precipitated when complicating diseases are present.

**Acknowledgment**

The author is deeply grateful to the Memorial Diagnostic Heart Service Association, Kansas City, Missouri, for the data of the initial right heart catheterization presented in case 1.

**References**


10. LENDRUM, B. L., AND SHAFFER, A. B.: Isolated...

On Permanent Patency of the Mouth of the Aorta, or Inadequacy of the Aortic Valves
By DOMINIC JOHN CORRIGAN, M.D.

One of the Physicians to the Charitable Infirmary, Jervis Street, Dublin; Lecturer on the Theory and Practice of Medicine; Consulting Physician to St. Patrick's College, Maynooth.

That this visible pulsation of the arteries is owing to the mechanical cause here assigned is made evident by several circumstances. It is most distinct in the arteries of the head and neck, which empty themselves most easily into the aorta, and of course into the ventricle. In the arteries of the lower extremities, of even larger size than those which present it about the head and neck, it is not seen to any comparative degree, and most generally not at all while the patient is standing or sitting. It is much more marked in the arteries of the head and neck in the erect than in the horizontal posture; and a patient suffering under the disease himself, first pointed out a circumstance which is convincing of its being produced as asserted. He could increase the pulsation of the brachial and palmar arteries in a most striking degree by merely elevating his arms to a perpendicular position above his head. He thus enabled the brachial and palmar arteries to empty themselves more easily back upon the aorta. They became more flaccid, and then, on the next contraction of the ventricle, their diastole became comparatively greater, and their visible pulsation of course more marked. The same effect could be produced in the arteries of the lower extremities by lying down and elevating the legs on an inclined plane.
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BEN O. PRICE

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