Combined Mitral and Pulmonic Stenosis

By Herbert Shubin, M.D., David C. Levinson, M.D., and Maurice H. Rosenfeld, M.D.

A CASE of coexisting stenosis of the mitral and pulmonic valves is described. Cardiac catheterization on a patient with stenosis of these 2 valves has not been reported to our knowledge. Gibson and Wood, however, described 1 case with coexisting stenosis of these 2 valves and the tricuspid valve as well.

A modification of the formula for calculating pulmonic valvular resistance is described.

Case Report and Methods

A 56-year-old Caucasian woman was well until the age of 25, when she had a full-term uncomplicated pregnancy. Several days following delivery, she first developed left precordial chest pain and was told by a physician that she had rheumatic heart disease with mitral stenosis. She had no history of rheumatic fever. The chest pain recurred intermittently for several days. She then became asymptomatic until the age of 32, when during the course of her second pregnancy, she first developed palpitation. The palpitation occurred infrequently at first, then with increasing frequency in the past 10 years, often 2 to 3 times per week with individual episodes lasting from several minutes to several days. On various occasions, they have been recorded by electrocardiogram as showing atrial tachycardia and atrial fibrillation. Quinidine was variably effective in terminating these paroxysmal attacks.

In April 1952, she had an initial episode of pulmonic edema. She was awakened from sleep with palpitation and dyspnea. Moist rales were present throughout both lung fields. She was treated with morphine sulfate and quinidine, and the pulmonary edema subsided as her heart rate slowed. An electrocardiogram taken after this episode showed first degree heart block (P-R interval of 0.22 second).

In April 1953, and again in March 1954, she had episodes of pulmonary edema arousing her from sleep and associated with marked tachycardia. As the cardiac rate returned to normal, the pulmonary edema subsided.

In December 1954, she had her first documented attack of paroxysmal atrial flutter, with the electrocardiogram showing a 2:1 atrial flutter. This arrhythmia was converted to a sinus rhythm with digitalis. Prior to this episode, she had not been treated with digitalis.

In July 1955, she had a paroxysm of atrial flutter that converted to atrial fibrillation while she was being digitalized, and subsequently to sinus rhythm with quinidine therapy. She has remained on digitalis since then, and has taken quinidine intermittently.

Aside from the periods of paroxysmal tachycardia, she has not had dyspnea on moderate exertion or undue fatigue. She has also been free of angina.

Physical examination disclosed a well developed woman in no discomfort. There was no cyanosis of the skin. The neck veins were not distended. The thyroid gland was not enlarged. The chest was symmetrical and moved well bilaterally. The lungs were clear to percussion and auscultation. The left lateral border of cardiac dullness extended to the fifth interspace in the midepigastric line. The point of maximum impulse and the apex beat were in the fifth interspace just medial to the midepigastric line. There was a diastolic thrill at the apex. The mitral first sound was loud. The pulmonic second sound was split and louder than the aortic second sound. An opening snap was heard at the left nipple area. A grade I blowing systolic murmur was heard at the pulmonic area. A grade I pansystolic rumble was present at the apex (fig. 1). The rhythm was atrial fibrillation with a ventricular rate of about 70. The blood pressure was 100/65 mm. Hg in the right arm and 112/72 mm. Hg in the right leg. Femoral pulsations were strong. The liver and spleen were not felt. The extremities showed no edema, clubbing, or deformity.

The hemoglobin was 11 Gm., blood count was 10,000, with the 70 per cent polymorphonuclear cells, 27 per cent lymphocytes, and 1 per cent each of eosinophils, basophils, and monocytes. The hematocrit level was 39 per cent. The urine was negative, the blood urea nitrogen was 12 mg. per 100 ml.

The electrocardiogram showed atrial fibrillation. The vectorcardiogram was normal.

Chest x-ray and fluoroscopy showed no enlargement of the heart. There was a double contour produced by moderate enlargement of the left atrium. There was a moderate increase in the angulation of the bifurcation of the trachea to 90° in the posteroanterior projection (normal less than 75°) and to a 65° angle in the left anterior oblique projection (normal less than 35°). There was slight posterior encroachment of the enlarged left atrium on the

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COMBINED MITRAL AND PULMONIC STENOSIS

Fig. 1. $S_1$, first heart sound; $S_2$, second heart sound; $OS$, opening snap; $DM$, diastolic murmur; $SM$, systolic murmur.

esophagus. The vascular markings in the lungs were normal (fig. 2).

Cardiac catheterization was performed in March 1956 (table 1). An abrupt pressure gradient of 13/8 to 38/8 mm. Hg was recorded as the catheter was withdrawn from the main pulmonary artery to the right ventricle (fig. 3), indicating a pulmonic valvular stenosis. The cardiac output of 1.6 L per minute was unusually low. There was significant venous oxygen desaturation and the A-V oxygen difference was great.

Calculations

Pulmonic Valvular Resistance. The method of determining pulmonic valvular resistance has been altered somewhat from that employed by Dow and co-workers who used the formula

$$PVR = \frac{(RV_{sm} - PA_{sm}) \times 1332}{CO \text{(ml./sec.)}} \text{ dynes seconds cm}^{-5}$$

$PVR = \text{Pulmonic valvular resistance. } RV_{sm} = \text{Right ventricular mean systolic ejection pressure in millimeters of mercury. } PA_{sm} = \text{Pulmonary artery mean systolic pressure in millimeters of mercury. } CO = \text{Cardiac output in milliliters per second = pulmonary blood flow in milliliters per second.}$

As Dow employed the formula, the flow past the pulmonic valve was computed as though it occurred throughout the cardiac cycle. Since the flow past the pulmonic valve occurs only during the systolic ejection phase of each cardiac cycle, the rate of flow per second is obtained by multiplying cardiac output (ml./sec.)

$$\times \frac{\text{duration of cardiac cycle}}{\text{duration of systolic ejection phase}}$$

Gorlin and Gorlin in calculating pulmonic valve area took note of this point, but did not employ this correction in calculating pulmonic valve resistance.

The formula for calculating pulmonic valve resistance should be

$$PVR = \frac{(RV_{sm} - PA_{sm}) \times 1332}{CO \text{(ml./sec.)} \times \text{duration of cardiac cycle} \times \text{duration of systolic ejection phase}} \text{ dynes seconds cm}^{-5}$$

The relation of the systolic ejection phase to the cardiac cycle depends on the heart rate. With increases in heart rate, the systolic ejection phase shortens, proportionately, than the diastolic phase, and therefore occupies a greater portion of each cardiac cycle. The effect of heart rate on the relative duration of the systolic ejection period is corrected in the revised formula.

Stenosis of the valve itself, as has been demonstrated with aortic stenosis, may also prolong the ejection phase of the cardiac cycle. The degree of stenosis may be an additional factor, therefore, in determining the relationship of the systolic ejection period to the total cardiac cycle. The effect of stenosis on the length of the systolic ejection phase is corrected in the revised formula.

In this case, each systolic ejection phase averaged about 0.24 second, and each cardiac cycle about 0.86 second, although with atrial fibrillation, this represents only an average value. The rate of flow past the pulmonic valve was therefore $0.86 = 3.57$ times $0.24$ as great as would be suggested by the cardiac output. The right ventricular mean systolic ejection pressure was 25 mm. Hg and the pulmonary artery mean systolic pressure was 10 mm. Hg. The pulmonic valvular resistance, therefore, was 210 dynes seconds cm$^{-5}$. Based on this method, normal resistance at the pulmonic valve would be expected to be less than 90 dynes seconds cm$^{-5}$. This calculation would be based on a cardiac output of 3 L. per minute or greater, a heart rate of about 70, and a difference in mean systolic pressure between the right ventricle and pulmonary artery of 10 mm. Hg or less. With a greater cardiac output per minute, or a smaller difference in mean systolic pressures between the right ventricle and pulmonary artery, the normal resistance at the pulmonic valve would
The arrows on the posteroanterior x-ray of the chest point out the double contour produced by the moderate enlargement of the left atrium. The lateral view of the chest shows slight posterior encroachment of the enlarged left atrium on the esophagus.

![Image](http://circ.ahajournals.org/)

**Fig. 2.**

**Fig. 3.** MPA, main pulmonary artery; PV, pulmonary valve; RV, right ventricle. The pressure changes abruptly between the main pulmonary artery and the right ventricle.

Pulmonic Valve Area. The area of the pulmonic valve may be derived from the formula by Gorlin and Gorlin, where

\[
PVA = \frac{PVF}{44.5V/\text{stim} - PA_s}
\]

where

- \(PVA\) = Pulmonic valve area in cm\(^2\)
- \(PVF\) = Pulmonic valve flow in milliliters per second (i.e., cardiac output in ml/sec. \(\times\) duration of cardiac cycle / duration of systolic ejection phase)
- \(RV_s\) = Right ventricular mean systolic ejection pressure in mm Hg
- \(PA_s\) = Pulmonary arterial mean systolic pressure in mm Hg

The pulmonic valve area in this patient is therefore 0.55 cm\(^2\), indicating a considerable degree of stenosis.

Pulmonary Vascular Resistance. The pulmonary vascular resistance proximal to the capillaries may be calculated by the formula

\[
PVR = \frac{(PA_m - PC_m) \times 1332}{CO (\text{ml/sec.})}
\]

where

- \(PVR\) = Pulmonary vascular resistance
- \(PA_m\) = Pulmonary artery mean pressure
- \(PC_m\) = Peripheral pulmonary artery (i.e., wedged or capillary) mean pressure
- \(CO\) = Cardiac output

The patient had a mean pulmonary artery pressure of 9 mm Hg and a mean peripheral pulmonary artery pressure of 4 mm Hg. Her pulmonic vascular resistance was 250 dynes seconds cm\(^{-5}\). Wood\(^4\) has calculated pulmonary vascular resistance in terms of units, where 1 unit represents approximately 80 dynes. By Wood's formula,

\[
PVR = \frac{PA_m - PC_m}{CO (L/min.)}
\]
TABLE 1.—Data Obtained during Cardiac Catheterization

<table>
<thead>
<tr>
<th>Station</th>
<th>Pressure (mm. Hg)</th>
<th>O2 saturation (vols. per cent)</th>
<th>Per cent O2 saturation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Superior vena cava</td>
<td>10/8</td>
<td>4.7</td>
<td>31.1</td>
</tr>
<tr>
<td>Right atrium</td>
<td>10/8</td>
<td>5.3</td>
<td>35.0</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>38/8</td>
<td>6.3</td>
<td>42.0</td>
</tr>
<tr>
<td>Main pulmonary artery</td>
<td>13/8</td>
<td>6.3</td>
<td>42.0</td>
</tr>
<tr>
<td>Right peripheral artery</td>
<td>5/3</td>
<td>14.7</td>
<td>97.5</td>
</tr>
</tbody>
</table>

O2 capacity = 15.1 vols. per cent = 100 per cent saturation. O2 consumption = 133 ml. per minute. Cardiac output = 1.6 L./min. (Fick principle). Cardiac index = 1.1 L./M.2/min.

By this formula the patient has a pulmonary vascular resistance of 3.1 units. Wood4 classified pulmonary vascular resistance of 3.9 units or less as normal, and resistance of over 10 units as extreme. He found that pulmonary vascular resistance of more than 10 units would usually protect against the development of pulmonary vascular congestion and dyspnea. Normal pulmonary vascular resistance of 3.9 units or less did not afford this protection. In this case, although the patient had a normal pulmonary vascular resistance of 3.1 units, she did not develop dyspnea on effort, except on those occasions when the dyspnea was brought on by paroxysmal tachycardia. In the presence of a normal vascular resistance, it is considered that the pulmonic stenosis protected her against pulmonary venous congestion and increasing pulmonary capillary pressure.

DISCUSSION

The cardiac output of 1.6 L. per minute is striking. Such a low output might be due to stenosis of either the mitral or pulmonic valves, or both. With so low an output due to mitral stenosis, greatly elevated peripheral pulmonary artery pressure would be expected but the peripheral pulmonary artery pressure was 5/3 mm. Hg. This value would indicate that the pulmonic stenosis exerted a significant limitation on right ventricular output. Both the pulmonic and mitral stenosis must have allowed for some increase in cardiac output under certain circumstances, however, for the moderate exercise that the patient was able to perform would hardly be possible with such a low cardiac output. Despite the low cardiac output she was not in cardiac failure. On 3 isolated occasions associated with paroxysmal tachycardia, she did have transient pulmonary edema. During innumerable other episodes of tachycardia, lasting often up to several days, she did not develop pulmonary edema. During these episodes of tachycardia, the pulmonic stenosis was sufficient, apparently, to prevent right ventricular output from being unduly greater than that of the left.

Mechanical limitation of right ventricular output, other than by pulmonic stenosis, is possible at the tricuspid valve, or by decreased venous return from the body. With marked tricuspid stenosis in the presence of mitral stenosis limitation of cardiac output might also occur at either or both of these valves, depending on the relative stenosis of each. In Gibson’s and Wood’s5 series of tricuspid stenoses, both low and normal cardiac outputs were recorded. It is a paradox, however, that in their series, the largest cardiac output (6.8 L. per minute) was recorded in the presence, not only of tricuspid and mitral stenosis, but of a third stenosis, pulmonic as well, indicating that the degree of functional stenosis is the governing factor. Limb tourniquets, like significant tricuspid stenosis, also may limit return to the right ventricle and right ventricular output.

SUMMARY

A case of combined pulmonic and mitral stenosis with cardiac catheterization studies is presented. A modification of the formula for calculating pulmonic valvular resistance is discussed. The importance of calculating cardiac output on the basis of the systolic ejection phase is noted.

The pulmonic valvular resistance by this modified formula was 210 dynes seconds cm.−5 The pulmonary vascular resistance was 250 dynes seconds cm.−5 The pulmonic valvular resistance tended to protect against the development of pulmonary edema, much as a high pulmonary vascular resistance protects against pulmonary edema.

The right ventricular output, which was 1.6 L. per minute, was limited by the pulmonic stenosis. Other mechanisms that might mechanically limit right ventricular output are signifi-
tricuspid stenosis or limb tourniquets. In situations of impending pulmonary edema, any one of these factors that decrease right ventricular output may be advantageous.

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Summario in Interlingua

Es presentate un caso de combine stenosis pulmonic e mitral, con studios de catheterisation cardia. Un modification del formula pro le calculation del resistantia del valvula pulmonic es discutite. Es signalate le importantia de calcular le rendimento cardia super le base del phase de ejection systolic.

Secundo le formula modificite, le resistantia pulmo-valvular eseva 210 dyn/sec/cm^2. Le resistantia pulmo-vascular eseva 250 dyn/sec/cm^2. Le resistantia pulmo-valvular exerceva un effecto protectori contra le disveloppamento de edema pulmonar, in plus o minus le mesme manera como alte resistantias pulmo-vascular es un protection contra edema pulmonar.

Le rendimento dextero-ventricular amontava a 1,6 l per minuta. Illo eseva limitate per le stenosis pulmonic. Altere mechanismos que es possibilemente capace a limitar le rendimento dextero-ventricular es grades significative de stenosis tricuspid e tourniquets al extremidades. In situationes de imminente edema pulmonar, non importa le qual de iste factores que reduce le rendimento dextero-ventricular es possibilemente avantageose.

References


Adams-Stokes Syndrome. Under these circumstances there was produced a group of symptoms and signs having a special character, namely, the combination of slow pulse, pseudo-apoplectic attacks, and murmur propagated into the aorta, while the second sound remained clear. In fact, my observations were based upon, and intended to illustrate, the views of Dr. Adams on fatty degeneration of the heart.—William Stokes. The Diseases of the Heart and the Aorta. Dublin, 1854.
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