Mitral Incompetence Caused by Disease of the Mural Cusp


Mitral regurgitation can be caused by one or a combination of different anatomic lesions. Because of this the authors believe that the surgeon can be aided if the type of defect is known preoperatively. The present study of 240 patients with disease of the mitral valve revealed a significant number whose disease was limited to the mural cusp. Criteria for the bedside diagnosis of this type of mitral regurgitation are presented.

Within the past few years there have been several reports of surgical procedures for the correction of mitral regurgitation. When, however, one studies the autopsy specimens of patients who had rheumatic mitral valve incompetence, it seems unlikely that any one operation short of complete replacement of the mitral valve would adequately repair the variety of anatomic defects that can exist. Brock, has investigated the anatomy of these lesions and has described shortening of one or both mitral cusps, rupture or shortening of the musculotendinous mechanism, stiffness of the cusp margins preventing closure, and dilatation of the atrioventricular ring. If the surgeon could be relatively certain which of the anatomic lesions was present, he could better determine the operative procedure best suited to correct that defect. The present study is an attempt to establish criteria for the diagnosis of one form of mitral incompetence.

When rheumatic valvulitis primarily affects the mural leaflet, it can reduce the size or mobility of the cusp in such a way that the relatively healthy aortic leaflet cannot close against it in systole. We believe this form of mitral regurgitation has specific diagnostic clinical features that can be recognized at the bedside.

Materials and Methods

Two hundred and forty patients with mitral valve disease were examined by one of us (P.N.) in the General Infirmary at Leeds during 1957 and 1958. Twenty-nine formed a distinct group, having in common the unusual combination of mitral incompetence with distinct mitral closing and opening snaps. In some of these 29 patients the incompetence was extreme, and opening and closing snaps persisted in the presence of left ventricular hypertrophy and the third heart sound of rapid early diastolic ventricular filling. In others, adhesion between the diseased mural cusp and the relatively healthy aortic leaflet had so reduced the size of the orifice that regurgitation appeared to be less important than stenosis and was clinically evident only in a pansystolic apical murmur. Eleven of these patients were sufficiently disabled to necessitate valvulotomy (G.H.W. and I.R.R.), which made digital examination of the mitral valve possible.

In each, the aortic cusp was found to be relatively healthy, but the mural cusp was diseased and permitted regurgitation. The operators' descriptions of the mitral valve in this group of 11 cases are listed in table 1. The important clinical and hemodynamic features observed in these same patients are tabulated in table 2, and one patient is reported in detail. Systemic blood pressures were obtained with a sphygmomanometer in patients supine and resting. The electrocardiographic criteria for the diagnosis of ventricular hypertrophy were those of Sokolow and Lyon. A phonocardiograph with a logarithmic frequency response was used to record sounds from the pulmonary area and apex.

Left atrial and pulmonary artery pressure pulses were obtained by needle puncture through a bronchoscope, using a modification of the technic devised by Allison and Linden. The zero reference point used was the sternal angle. All records were taken during an expiratory pause.

Case Report

Case 8, E.H., female, age 42 years. Apart from chorea at 13 years, there was no illness until 1943, when breathlessness on effort and fatigue were first noticed. The severity of the symptoms altered little until early in 1956, when dyspnea grew troublesome. In the summer of 1957 she had to
give up work because traveling and climbing stairs had become difficult. At the time of the present examination, in July 1958, dyspnea and fatigue prevented her from walking more than 25 yards without resting. It was necessary for her to sleep propped up in bed. There had been no hemoptysis and no edema. There was no evidence of disease other than the cardiac disorder.

Examination showed a tall lean patient with malar cyanosis and cold, blue hands but no central cyanosis. The jugular venous pressure was normal. There was no enlargement of the liver or edema. The radial pulse was irregular but of normal quality. The blood pressure was 140/90. Her apex beat was palpable in the seventh left intercostal space in the midaxillary line. The cardiac impulse was a well-marked apical thrust of left ventricular hypertrophy, and a parasternal heave of right ventricular enlargement. The pulmonary valve closure was easily palpable. The mitral first sound was accentuated and followed by a loud pansystolic murmur, second sound, opening snap, third sound, and short diastolic murmur. A quiet pansystolic murmur at the left sternal border was considered to be conducted from the mitral area. The second sound in the pulmonary area split normally during inspiration, and the sound of pulmonary valve closure was accentuated. There was no evidence of aortic valvular disease.

The lungs, abdomen, and nervous system were normal. Blood count and sedimentation rate were normal.

The electrocardiogram showed atrial fibrillation, vertical heart position, and left ventricular hypertrophy (fig. 1). The phonocardiogram confirmed the auscultatory findings (fig. 2). X-ray examination (fig. 3) showed enlargement of the ventricles, small aortic knuckle, prominence of the hilar, vessels, and pulmonary venous congestion. Horizontal lines were probably present, but not distinct. The left atrium was markedly enlarged, and formed part of the right border of the heart in the posteroanterior film. Eighteen months before the present examination the left atrial and pulmonary artery pressures (fig. 4) were recorded: Pulmonary artery mean pressure, 28 mm. Hg; mean left atrial pressure, 15.5 mm. Hg; left atrial pressure at “v”, 28.4 mm. Hg; Ry/v factor, 3.6; V/M factor, 1.86.

Operation was performed on July 10, 1958. Using the phonocardiograph to provide an extra-corporal circulation of 1.7 L./M.2/minute at a body temperature of 33.5 C., the left atrium was opened widely and the mitral valve examined. The lumen measured 2.5 cm. in diameter. The aortic leaflet was mobile and only slightly thickened. The mural cusp was thickened, rolled upon itself, and tethered down by shortened chordae.

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**Table 1.—Condition of the Mitral Valve at Cardiomyotomy**

<table>
<thead>
<tr>
<th>Case</th>
<th>Remarks</th>
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</thead>
<tbody>
<tr>
<td>1 W.F.</td>
<td>Orifice approximately 2 em. in diameter. Posterior commissure thickly fibrosed. Aortic leaflet reasonably healthy, and moving a great deal. Mural cusp immobile and thickened. Regurgitant stream increased by division of the posterior commissure.</td>
</tr>
<tr>
<td>2 I.S.</td>
<td>Orifice approximately 1.5 cm. in diameter. Aortic leaflet mobile. Mural cusp completely fixed. Regurgitant jet increased by division of the anterior commissure.</td>
</tr>
<tr>
<td>3 K.S.</td>
<td>Orifice approximately 3.0 cm. in diameter. Gross regurgitant stream. Mural cusp thickened, rolled upon itself, and immobile. Aortic cusp appeared undiseased and mobile, but could not meet the retracted mural cusp.</td>
</tr>
<tr>
<td>4 E.M.</td>
<td>Orifice approximately 2.5 cm. in diameter. Gross regurgitant stream. Aortic leaflet mobile, but unable to meet the diseased mural cusp.</td>
</tr>
<tr>
<td>5 H.L.</td>
<td>Orifice approximately 1.2 cm. in diameter. Regurgitant jet. Aortic leaflet almost undiseased and freely mobile. Mural cusp grossly diseased, thickened, and immobile.</td>
</tr>
<tr>
<td>6 M.J.</td>
<td>Orifice approximately 2.5 cm. in diameter. Regurgitant stream. Aortic leaflet hardly diseased and very mobile. Mural cusp immobile, thickened, and rolled upon itself so that the aortic leaflet could not meet it.</td>
</tr>
<tr>
<td>7 M.L.</td>
<td>Orifice approximately 3 cm. in diameter. Regurgitant jet. Aortic leaflet freely mobile and apparently undiseased. Mural cusp greatly thickened and rolled upon itself so that it could not be met by the aortic leaflet.</td>
</tr>
<tr>
<td>8 E.H.</td>
<td>Orifice approximately 2.5 cm. in diameter. Considerable regurgitant flow. Aortic leaflet pliant and mobile. Mural cusp thickened, rolled, immobile, and held to the ventricular wall by short thick chordae tendineae.</td>
</tr>
<tr>
<td>9 L.S.</td>
<td>Orifice approximately 1.5 cm. in diameter. Moderate regurgitant jet. Aortic leaflet full and mobile. Mural cusp rigid and immobile.</td>
</tr>
<tr>
<td>10 W.P.</td>
<td>Orifice approximately 2 cm. in diameter. Regurgitant stream. Aortic cusp pliant and mobile. Mural cusp shrunved and functionless.</td>
</tr>
<tr>
<td>11 A.B.</td>
<td>Orifice approximately 1.5 cm. in diameter. Regurgitant jet. Aortic cusp freely movable and ballooned normally. Its edge was thickened and fused at the lateral and medial commissures to a thickened, narrow, and immobile mural cusp.</td>
</tr>
</tbody>
</table>
MITRAL INCOMPETENCE CAUSED BY DISEASE OF MURAL CUSP

Table 2.—Cardiographic and Hemodynamic Findings in Eleven Patients with Mural Cusp Disease

<table>
<thead>
<tr>
<th>Patient</th>
<th>Rhythm</th>
<th>Mitral P wave</th>
<th>Electrical position of heart</th>
<th>Ventricular hypertropy</th>
<th>Cardiographic</th>
<th>Mean left atrial pressure (mm. Hg)</th>
<th>Left atrial pressure at “V” (mm. Hg)</th>
<th>Mean pulmonary artery pressure (mm. Hg)</th>
<th>Systemic blood pressure (mm. Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 W.P.</td>
<td>SR</td>
<td>X</td>
<td>Vertical</td>
<td>Left</td>
<td>Cardiographic</td>
<td>16</td>
<td>21</td>
<td>1.44</td>
<td>21</td>
</tr>
<tr>
<td>2 I.S.</td>
<td>SR</td>
<td>X</td>
<td>Vertical</td>
<td>?</td>
<td>Cardiographic</td>
<td>15</td>
<td>22</td>
<td>3</td>
<td>32</td>
</tr>
<tr>
<td>3 K.S.</td>
<td>AF</td>
<td>—</td>
<td>Vertical</td>
<td>Left</td>
<td>Cardiographic</td>
<td>17</td>
<td>27.5</td>
<td>3.55</td>
<td>44</td>
</tr>
<tr>
<td>4 E.M.</td>
<td>SR</td>
<td>X</td>
<td>Vertical</td>
<td>Right</td>
<td>Cardiographic</td>
<td>33</td>
<td>45.5</td>
<td>2.25</td>
<td>60</td>
</tr>
<tr>
<td>5 H.L.</td>
<td>AF</td>
<td>—</td>
<td>Vertical</td>
<td>Left</td>
<td>Cardiographic</td>
<td>30</td>
<td>41.4</td>
<td>1.18</td>
<td>40</td>
</tr>
<tr>
<td>6 M.J.</td>
<td>AF</td>
<td>—</td>
<td>Intermediate</td>
<td>Left</td>
<td>Cardiographic</td>
<td>18.4</td>
<td>22</td>
<td>2.64</td>
<td>30</td>
</tr>
<tr>
<td>7 M.I.</td>
<td>AF</td>
<td>—</td>
<td>Vertical</td>
<td>None</td>
<td>Cardiographic</td>
<td>20.6</td>
<td>28</td>
<td>0.95</td>
<td>—</td>
</tr>
<tr>
<td>8 E.H.</td>
<td>AF</td>
<td>—</td>
<td>Vertical</td>
<td>Left</td>
<td>Cardiographic</td>
<td>15.5</td>
<td>28.5</td>
<td>3.6</td>
<td>28</td>
</tr>
<tr>
<td>9 L.S.</td>
<td>SR</td>
<td>X</td>
<td>Vertical</td>
<td>None</td>
<td>Cardiographic</td>
<td>19.5</td>
<td>25</td>
<td>3.8</td>
<td>—</td>
</tr>
<tr>
<td>10 W.F.</td>
<td>SR</td>
<td>X</td>
<td>Intermediate</td>
<td>None</td>
<td>Cardiographic</td>
<td>—</td>
<td>—</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>11 A.B.</td>
<td>SR</td>
<td>X</td>
<td>Vertical</td>
<td>None</td>
<td>Cardiographic</td>
<td>22</td>
<td>40</td>
<td>1.65</td>
<td>—</td>
</tr>
</tbody>
</table>

*Owen and Wood.*

![Fig. 1. Case 8, electrocardiogram.](http://circ.ahajournals.org/)

Tendineae. Regurgitation occurred through the central and posterior parts of the lumen. The anterior commissure was obliterated. The anterior commissure was opened with scissors. Posterior regurgitation was reduced by the insertion of 2 sutures through the posterior part of the annulus. A piece of Ivalon sponge was then sutured to the mural cusp to form a cushion against which the aortic leaflet could close. This patient made an uneventful recovery.

**DISCUSSION**

The patients in this study were carefully examined with particular reference to the physical signs of mitral closing and opening snaps, the pansystolic murmur of mitral re-
gurgitation, the third heart sound of rapid early diastolic ventricular filling, and the thrusting cardiac impulse of left ventricular hypertrophy. The criteria for the recognition of these signs are well established and have been reviewed by Wood. In the cases we report, the closing snap or loud mitral first sound has contrasted strongly with the usually quiet first heart sound of mitral incompetence. By deduction it would thus seem that a quiet first sound would indicate fixed or destroyed mitral cusps.

The systolic murmur has been easily heard and in our patients has not been simulated by the ejection murmur of aortic stenosis. The only difficulty we have encountered has occurred in distinguishing between the pansystolic murmurs of mitral and tricuspid regurgitation in those patients with little or no left ventricular hypertrophy and in whom right ventricular enlargement has caused clockwise rotation of the heart.

The opening snap was readily audible in 10 of the 11 patients who were operated upon. In one it was recognized with difficulty because the mitral incompetence had shortened ventricular systole to such a degree that mitral valve opening nearly coincided with the loud pulmonary valve closure. The third heart sound of rapid early diastolic ventricular filling must occur after the opening snap. Unlike the opening snap, this sound is of a low frequency and can rarely be heard at the pulmonary area. While it has been possible to record the third sound by phonocardiography, we have had difficulty in distinguishing it by auscultation within the short, loud, almost explosive diastolic murmur that has been present in some cases of severe mitral incompetence.

Causation of Mitral Opening and Closing Snaps

The importance of cusp mobility in the production of mitral opening and closing snaps was recognized when advances in cardiac surgery stimulated a close examination of the meaning of the physical signs in mitral valve disease. Prior to this, it was considered that the presence of these snaps excluded gross incompetence. Recent reports, however, have described the occurrence of opening and closing snaps in patients with dominant regurgitation. In addition, Wood and McDonald et al. have heard the third sound of rapid early diastolic ventricular filling in patients with opening snaps.
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All the patients who were operated upon in this series had mitral incompetence and, in some, this was the dominant lesion. All of them had opening or closing snaps, or both, and none of them had a mobile or effective mural cusp. It therefore seems clear that their opening and closing snaps must be caused by upward and downward movement of a mobile aortic cusp.

Possible Clinical Course of Mural Cusp Disease and Surgical Considerations

The mural cusp normally seals only a narrow crescent of the mitral orifice; so it is probable that regurgitation is slight in the early stages of its disease. Burchell and Edwards have speculated upon the consequences of regurgitation in a case of the type we describe, and suggest that it leads to dilatation of the left atrium, which in turn causes posterior retraction of the mural cusp and increased regurgitation, and sets up a vicious circle that ends in gross incompetence and marked dilatation of the atrioventricular ring. If this view is correct it follows that there may be advantage in treating mural cusp disease when the defect is small and not too difficult to close with a prosthesis. We consider that there may be a second reason for operating at a relatively early stage of this disease, namely, the possibility that commissurotomy to free the aortic cusp may greatly increase regurgitant flow if dilatation of the atrioventricular ring has already occurred.

An approach to the treatment of mural cusp disease has already been made. Sakakariba employed a sling to elevate the edge of the mural cusp after freeing the aortic cusp by commissurotomy. Making use of advances in the technic of extracorporeal circulation, Lillehei et al., and the present authors have sutured Ivalon sponge to the mural cusp to form a cushion against which the pliant and mobile aortic cusp can close in systole.

Incidence of Mural Cusp Disease

Only a relative idea of the incidence of mural cusp disease can be drawn at present. The characteristic physical signs were found in 29 (12 per cent) of a series of 240 patients with mitral valve disease examined by one observer in the Cardiological and Thoracic Surgical Clinics of a general hospital during 1957 and 1958. Most cases were referred for consideration of mitral valvulotomy and 120 had this operation performed. Only those patients in whom mitral valve disorder was the chief cardiac lesion were included in the series.

Summary

A syndrome that has received little attention in the literature was present in 12 per cent of a series of 240 cases of mitral valve disease. The characteristic clinical feature was the presence of mitral opening and closing snaps in patients with mitral incompetence. The valve was examined at cardiotomy in the 11 cases reported, and the cause of regurgitation in each was disease of the mural cusp, the aortic leaflet being pliant and mobile.

Since none of the cases reported had an effective mural cusp, and since some had considerable regurgitation, it follows that mitral opening and closing snaps are produced by movement of the aortic leaflet.

It has been found possible at open heart operation to relieve the regurgitation of mural cusp disease by suturing a prosthesis to the mural cusp in such a way as to form a cushion against which the aortic leaflet can close in systole. It is suggested that the condition is best treated before serious dilatation of the mitral annulus occurs.

Acknowledgment

The authors wish to thank Dr. William Whitaker for his helpful advice and criticism.
Nixon, Wooler, Radigan


SUMMARIO IN INTERLINGUA

Un syndrome a que pauc attention es presente in le litteratura esseva inontrate in 12 pro cento de un serie de 240 casos de morbo del valvula mitral. Le characteristic aspecto clinic esseva le presentia de clics de apertura e clauditura in patientes con incompetentia mitral. In le 11 casos hic reportate, le valvula esseva examine in cardiotomia, e le causa del regurgitation, in omne iste casos, esseva morbo del cuspile mitral—con plicabilitate e mobilitate del folio aortie.

Viste que in nulle del casos hic reportate il habeva un efficace cuspile mitral e que in certes il habeva considerable grados de regurgitation, il seque que le clics de apertura e clauditura mitral es producete per movimientos del folio aortie.

Il se ha provate possibile alleviar, in operaciones cardiac aperte, le regurgitation de morbo de cuspile mitral per suturar un prosthes al cuspile mitral de maniera a formar un cossino contra le qual le folio aortie poto effectuar un clauditura in systole. Es expressite le opinion que le condition es a tractar preferibilmente ante que un serie dilatation del anulo mitral ha occurrute.

REFERENCES


3. — AND —: The ventricular complex in left
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