Combined mitral and aortic valve stenosis caused by two different etiologies, rheumatic and congenital

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Table 1. Pertinent findings in the five patients with stenotic congenitally bicuspid aortic valves and stenotic rheumatic mitral valves

<table>
<thead>
<tr>
<th>Case number</th>
<th>Age at VR (years)</th>
<th>Sex</th>
<th>BMI (kg/m²)</th>
<th>PAW (mm Hg)</th>
<th>RV (s/d)</th>
<th>RA (s/d)</th>
<th>LV (mm Hg)</th>
<th>Ao (mm Hg)</th>
<th>LV-Ao (mm Hg)</th>
<th>AV area (cm²)</th>
<th>MV area (cm²)</th>
<th>AV weight (g)</th>
<th>MV weight (g)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>44</td>
<td>M</td>
<td>26</td>
<td>21</td>
<td>86/19</td>
<td>16</td>
<td>126/14</td>
<td>108/66</td>
<td>18</td>
<td>16</td>
<td>–</td>
<td>1.54</td>
<td>0.9</td>
</tr>
<tr>
<td>2</td>
<td>46</td>
<td>M</td>
<td>26</td>
<td>26</td>
<td>48/18</td>
<td>13</td>
<td>139/17</td>
<td>133/88</td>
<td>28</td>
<td>15</td>
<td>0.46</td>
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<td>1.37</td>
</tr>
<tr>
<td>3</td>
<td>50</td>
<td>M</td>
<td>27</td>
<td>39</td>
<td>56/18</td>
<td>13</td>
<td>173/40</td>
<td>146/88</td>
<td>27</td>
<td>25</td>
<td>1.0</td>
<td>1.28</td>
<td>–</td>
</tr>
<tr>
<td>4</td>
<td>53</td>
<td>F</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>–</td>
<td>0.9</td>
<td>1.8</td>
<td>–</td>
</tr>
<tr>
<td>5</td>
<td>74</td>
<td>F</td>
<td>21</td>
<td>33</td>
<td>79/17</td>
<td>54</td>
<td>226/19</td>
<td>195/85</td>
<td>31</td>
<td>–</td>
<td>0.53</td>
<td>0.95</td>
<td>–</td>
</tr>
</tbody>
</table>

Ao indicates aorta; AV, aortic valve; BMI, body mass index; LV, left ventricular; mdg, mean diastolic gradient; MV, mitral valve; PAW, pulmonary artery wedge; psg, peak systolic gradient; RA, right atrium; RV, right ventricular; s/d, peak systole/end diastole; VR, valve replacement.

Described herein are five patients who had double left-sided cardiac valve replacement for mitral and aortic valve stenosis resulting from two different etiologies: rheumatic heart disease, the cause of the mitral stenosis, and congenital heart disease (bicuspid valve), the underlying cause of the aortic stenosis.

Combined mitral and aortic valve stenosis in the same patient today in the Western world is relatively uncommon. Worldwide, its most common cause is rheumatic heart disease. Today in native Europeans and in European Americans, the most common cause of this combination appears to be massive mitral annular calcification (probably the consequence of atherosclerosis) as the cause of the mitral stenosis and either congenital (unicuspid or bicuspid aortic valve) or nonrheumatic causes (tricuspid aortic valve) as the cause of the aortic stenosis (atherosclerotic etiology) (1). On very rare occasions, the mitral stenosis may be the result of rheumatic heart disease and the aortic stenosis, the result of an underlying congenital heart disease (unicuspid or bicuspid aortic valve). Such was the case in the five patients described herein.

METHODS

Shown in Table 1 are pertinent findings in the five patients with combined rheumatic mitral stenosis and aortic stenosis superimposed on a congenitally bicuspid aortic valve. Photographs of the operatively excised valves are shown in Figures 1 and 2. Only Case 1 is still alive. Cases 2 to 5 survived from 6.9 to 14.4 years (mean 11.2) after the double valve replacement.

DISCUSSION

The patients described here had two different causes for the two stenotic valves on the left side of the heart: rheumatic heart disease for the mitral stenosis and congenital heart disease (bicuspid aortic valve) for the aortic stenosis. Relatively few such combinations have been reported previously. McReynolds and associates (2) in 1976 described an 18-year-old man with a purely regurgitant congenitally bicuspid aortic valve (without commissural fusion) and a diffusely fibrotic nonstenotic mitral valve proven to be of rheumatic etiology by the presence of Aschoff nodules in the left atrial wall. Tejada and associates (3) in 2001 described a 60-year-old woman who had a noncalcified congenitally bicuspid aortic valve and percutaneous valvuloplasty for a typical rheumatic stenotic mitral valve. Muthiah (4) in India in 2016 described by echocardiogram a

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41-year-old man with a “congenitally malformed” stenotic aortic valve and a stenotic mitral valve that appeared to be of rheumatic origin.

On occasion, in patients with rheumatic mitral stenosis, the aortic valve may have fusion of one of its three commissures, simulating a congenitally bicuspid aortic valve. In these circumstances, however, the ventricular aspect of the fusion appears as a V-shaped indentation, producing some space below the fusion. In contrast, with a congenitally bicuspid aortic valve, the ventricular aspect of the raphe area, which some may interpret as fusion of the two cusps, is flat, such that there is no space beneath the fusion. Such was the case in each of the five patients described herein, indicating that the aortic valve in each was congenitally malformed and not the result of rheumatic heart disease.


Figure 1. Case 1. Mitral valve as viewed from the (a) ventricular aspect and (b) atrial aspect. Aortic valve as viewed from the (c) aortic aspect and (d) ventricular aspect.

Figure 2. Case 2. (a) Thickened mitral valve and (b) bicuspid aortic valve.