Parachute mitral valve: Morphology and surgical management
Paraşüt mitral kapak: Morfoloji ve cerrahi tedavi

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ABSTRACT

Background: This review aims to discuss morphology and surgical management of parachute mitral valve.

Methods: A total of 62 articles in the English language with 330 parachute mitral valve patients were retrieved from the PubMed, HighWire Press, and Cochrane Library databases using specific MeSH terms and keywords between January 2000 and December 2018. In these articles, morphology of parachute mitral valve and surgical treatment options were investigated.

Results: A non-syndromic parachute mitral valve was present in 287 patients (87.0%) and a syndromic parachute mitral valve was present in 43 patients (13.0%). A higher number of patients with a non-syndromic parachute mitral valve presented with congestive heart failure compared to syndromic ones. The patients with a non-syndromic parachute mitral valve often had mitral regurgitation, while syndromic parachute mitral valve patients often had mitral stenosis.

Conclusion: Parachute mitral valves are usually not an isolated lesion and are often characterized by a constellation of pathological changes of the mitral valve leaflets, annulus, commissures, subvalvular apparatus, and supravalvular mitral ring. Therefore, the majority of the patients need one or more surgical operations. The incidence of adverse events such as reintervention, postoperative complete heart block, and mortality is high in these patients.

Keywords: Cardiac surgery; mitral valve annuloplasty; mitral valve stenosis.

In 1963, Shone et al. [1] firstly reported the mitral valve pathology of “an insertion of the chordae into a single papillary muscle, producing a funnel-shaped valve”, and they defined this lesion as a parachute mitral valve (PMV). Subsequently, Bett and Stovin [2] reported a patient with PMV and bicuspid aortic valve. In PMV, all chords are typically shortened and thickened, and attached to the posteromedial papillary muscle, while the anterolateral papillary muscle is absent. [3-5]
Parachute mitral valve can be an isolated lesion, or one of the constellations of Shone syndrome. McElhinney et al.\textsuperscript{[6]} reported that 25.9% patients with severe congenital mitral stenosis had a PMV. Aslam et al.\textsuperscript{[7]} also reported Shone syndrome in 1.17% of all congenital heart lesions. Shone syndrome mainly consists of four defects: supravalvular mitral membrane, PMV, subaortic stenosis (membranous or muscular), and coarctation of the aorta.\textsuperscript{[1,8]} The solitary papillary muscle and orientation of a severely affected PMV contributes to subaortic stenosis.\textsuperscript{[9]} Although surgical management of PMV is constantly reported, the morphological features of PMV and surgical indications of PMV have been described in limited cases.

In this review, pertinent morphological aspects and surgical management of PMV are discussed.

**MATERIALS AND METHODS**

Publications were systematically searched in the PubMed, HighWire Press, and Cochrane Library databases between January 2000 and December 2018. The MeSH terms and keywords were used to identify articles including “parachute mitral valve”, “supravalvular mitral ring”, “single papillary muscle”, “Shone syndrome”, “congenital mitral stenosis”, “mitral valve repair”, and “mitral valve replacement”, in the English language. The screening of the bibliographic references helped in completing the literature retrieval. A total of 62 articles including 17 retrospective studies, one case series, and 44 case reports which met the inclusion criteria during initial screening were included in this review. Double-blind, randomized-controlled clinical studies were excluded.

The data independently extracted from each publication were the patient demographics, clinical presentations, mitral valve morphology, cardiac surgical procedures, and patient outcomes.

**Statistical analysis**

Statistical analysis was performed using the IBM SPSS version 22.0 software (IBM Corp., Armonk, NY, USA). Descriptive data were expressed in mean ± standard deviation (SD), median (min-max) or number and frequency. Independent samples t-test was used to compare quantitative variables. The categorical variables were compared using the chi-square ($\chi^2$) or Fisher’s exact test with continuity correction. A $p$ value of <0.05 was considered statistically significant.

**RESULTS**

In total, 62 articles\textsuperscript{[3,5-7,9-66]} with 330 patients were included. A non-syndromic PMV was present in 287 (87.0%) patients and a syndromic PMV (as constellation of pathology of Shone syndrome) was present in 43 (13.0%) patients.

Gender was described for 236 (236/330 patients, 71.5%) and 135 (57.2%) were males and 101 (42.8%) were females. The male-to-female ratio did not differ between patients with non-syndromic and syndromic PMV (128/95 vs. 7/6, $\chi^2=0.1$, $p=1.000$). The mean age of the patients was 21.1±22.0 (range, 0 to 85) years. Age was not specified in 224 patients. A total of 166 (74.1%) patients were pediatric and 58 (25.9%) were adult patients ($\chi^2=104.1$, $p<0.001$). In the non-syndromic PMV group, 145 (145/177, 81.9%) patients were pediatrics and 32 (32/177, 18.1%) were adults, while in the syndromic PMV group, 21 (21/47, 44.7%) patients were pediatrics and 26 (26/47, 55.3%) patients were adults.

Most PMV patients were symptomatic, while a few patients were asymptomatic in either non-syndromic or syndromic PMV patients ($\chi^2=42.6$, $p<0.001$ for non-syndromic patients, and $\chi^2=16.7$, $p<0.001$ for syndromic patients). Among pediatrics, 17 patients (21.3%) were asymptomatic and 63 patients (78.8%) were symptomatic, while among adults, nine patients (36%) were asymptomatic and 16 patients (64%) were symptomatic ($\chi^2=2.2$, $p=0.184$). A higher number of patients with a non-syndromic PMV presented with congestive heart failure compared to syndromic ones. The patients with a non-syndromic PMV were mostly associated with an atroventricular septal defect and hypoplastic left ventricle than those with a syndromic PMV. However, the patients with a syndromic PMV had a higher incidence of coarctation of the aorta, bicuspid aortic valve, and subaortic obstruction (Table 1).

Hemodynamic studies showed that the peak and mean mitral pressure gradients did not significantly differ between the groups (Table 1). All patients had a single papillary muscle. Most patients had a thickened mitral valve leaflet, shortened chords, and mitral stenosis or regurgitation. The patients with a non-syndromic PMV often had mitral regurgitation, while those with a syndromic PMV had mitral stenosis (Table 2).

Management was described for 216 patients. Accordingly, 20 patients (9.3%) were not operated due to conservative treatment/on a follow-up/waiting for surgical operation/operation refusal/sudden death, and loss of surgical opportunity.\textsuperscript{[12,13,15,21,22,25,26,29,41,42,45,48-52,57,66]} while 196 patients (90.7%) received a total of 198 surgical operations as follows: non-mitral valve operations (n=70, 35.4%)\textsuperscript{[3,11,14,26,30,33,36,39,46,61]} and
### Table 1. Comparisons between non-syndromic and syndromic PMV patients

<table>
<thead>
<tr>
<th>Clinical presentation</th>
<th>Non-syndromic PMV</th>
<th>Syndromic PMV</th>
<th>Test</th>
<th>(\chi^2)</th>
<th>(p)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Case number</td>
<td>287 87.0</td>
<td>43 13.0</td>
<td>Chi-square</td>
<td>360.8</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Gender</td>
<td></td>
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</tr>
<tr>
<td>Male (n=135)</td>
<td>128 94.8</td>
<td>7 5.2</td>
<td>Chi-square</td>
<td>0.1</td>
<td>1.000</td>
</tr>
<tr>
<td>Female (n=101)</td>
<td>95 94.1</td>
<td>6 5.9</td>
<td>Chi-square</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Child/adult</td>
<td></td>
<td></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Child (n=166)</td>
<td>145 87.3</td>
<td>21 12.7</td>
<td>Chi-square</td>
<td>26.8</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Adult (n=58)</td>
<td>32 55.2</td>
<td>26 44.8</td>
<td>Chi-square</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Asymptomatic (n=26)</td>
<td>25* 26.3</td>
<td>1 8.3</td>
<td>Fisher exact test with continuity correction</td>
<td>1.9</td>
<td>0.286</td>
</tr>
<tr>
<td>Symptomatic (n=81)</td>
<td>70 73.7</td>
<td>11 91.7</td>
<td>Chi-square</td>
<td>1.9</td>
<td></td>
</tr>
<tr>
<td>Heart failure</td>
<td>38 54.3</td>
<td>1 9.1</td>
<td>Fisher exact test with continuity correction</td>
<td>7.8</td>
<td>0.007</td>
</tr>
<tr>
<td>Dyspnea/tachypnea</td>
<td>14 20.0</td>
<td>8 72.7</td>
<td>Chi-square</td>
<td>13.4</td>
<td>0.001</td>
</tr>
<tr>
<td>Associated cardiac anomaly</td>
<td></td>
<td></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>Coarctation of the aorta</td>
<td>82 28.6</td>
<td>33 76.7</td>
<td>Chi-square</td>
<td>38.2</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Bicuspid aortic valve</td>
<td>63 22.0</td>
<td>30 69.8</td>
<td>Chi-square</td>
<td>68.3</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Hypoplastic left ventricle</td>
<td>36 12.5</td>
<td>0 0</td>
<td>Fisher exact test with continuity correction</td>
<td>6.1</td>
<td>0.008</td>
</tr>
<tr>
<td>Atrioventricular septal defect</td>
<td>26 9.1</td>
<td>0 0</td>
<td>Fisher exact test with continuity correction</td>
<td>4.2</td>
<td>0.034</td>
</tr>
<tr>
<td>Subaortic obstruction</td>
<td>25 8.7</td>
<td>12 27.9</td>
<td>Chi-square</td>
<td>13.8</td>
<td>0.001</td>
</tr>
<tr>
<td>Tricuspid regurgitation</td>
<td>3 1.0</td>
<td>4 9.3</td>
<td>Fisher exact test with continuity correction</td>
<td>12.3</td>
<td>0.007</td>
</tr>
<tr>
<td>Anomalous origin of coronary artery</td>
<td>1 0.3</td>
<td>2 4.7</td>
<td>Fisher exact test with continuity correction</td>
<td>7.7</td>
<td>0.046</td>
</tr>
<tr>
<td>Left superior vena cava</td>
<td>1 0.3</td>
<td>7 16.3</td>
<td>Fisher exact test with continuity correction</td>
<td>40.1</td>
<td>&lt;0.001</td>
</tr>
<tr>
<td>Supravalvular aortic stenosis</td>
<td>1 0.3</td>
<td>3 7.0</td>
<td>Fisher exact test with continuity correction</td>
<td>13.7</td>
<td>0.008</td>
</tr>
</tbody>
</table>

PMV: parachute mitral valve; * One of the asymptomatic patients developed dyspnea later.\(^{[41]}\)
Table 2. Hemodynamic, metrology, and morphology of PMV

<table>
<thead>
<tr>
<th>Clinical presentation</th>
<th>Non-syndromic PMV (n=287)</th>
<th>Syndromic PMV (n=43)</th>
<th>Test</th>
<th>( \chi^2 / t )-value</th>
<th>( P )</th>
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</thead>
<tbody>
<tr>
<td>Mean mitral pressure gradient (mmHg)</td>
<td></td>
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<td>0.763</td>
<td>0.458</td>
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<td>Peak mitral pressure gradient (mmHg)</td>
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<td></td>
<td></td>
<td>0.673</td>
<td>0.520</td>
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<td>Annulus diameter (mm)</td>
<td>11.6±4.4</td>
<td>21.5±11.0</td>
<td>Independent samples t test</td>
<td>0.352</td>
<td>0.732</td>
</tr>
<tr>
<td>Mitral valve area (cm(^2))</td>
<td>1.8±1.0</td>
<td>2.0±0.6</td>
<td>Independent samples t test</td>
<td></td>
<td></td>
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<tr>
<td>Dominant papillary muscle</td>
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<tr>
<td>• Posteromedial (n=41)(^{[39,40,45,48,53,62]})</td>
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<tr>
<td>• Anterolateral (n=14)(^{[39,65]})</td>
<td></td>
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<td></td>
<td></td>
<td></td>
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<tr>
<td>• Inferolateral(^{[56]})</td>
<td></td>
<td></td>
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<td></td>
<td></td>
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<tr>
<td>• Left (n=1)(^{[32]})</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
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<tr>
<td>• Central(^{[34]})</td>
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</table>

PMV: parachute mitral valve; MR: Mitral regurgitation; MS: Mitral stenosis.
mitral valve operations (n=128, 64.6%). Mitral valve operations were prevailed by double patch repair for atrioventricular septal defects (n=20, 15.6%).[44] Surgical mitral valvuloplasty (n=17, 13.3%)[5,66] mitral valve replacement (MVR) (n=16, 12.5%),[3,5,9,26,34,36,59,62,66] and zone of apposition closure (n=16, 12.5%).[58] Reintervention was required in 19 patients (19/196, 9.7%).[3,9,16,17,26,58,60]

The mean follow-up of the patients was 150.7±145.6 months (range, 1 month to 20 years) (n=21). The outcomes of 231 patients were reported as follows: 191 (82.7%) recovered, six (2.6%) were complicated, and 34 (14.7%) died.

**DISCUSSION**

**Morphology**

Parachute mitral valve is formed by specific malformations of the mitral leaflets per se, as well as subvalvular structures (Table 3). The major morphology of PMV is a single papillary muscle, or one papillary muscle is severely hypoplastic. Chauvaud[67] proposed that, in PMV patients, mitral regurgitation might be caused by hypoplasia of one papillary muscle, commissural enlargement, valve leaflet defects, and shortened chords. The authors reported that the hypoplastic papillary muscle was usually the posterior one, while the other papillary muscle was mediially displaced. However, some others[39,43,45,48,53,62] reported the dominant papillary muscle was the posterior one. The results of this study supported the posterior papillary muscle was the dominant one.

The combination of lesions can give rise to a funnel configuration of the mitral valve. Three-dimensional echocardiography can visualize all characteristic findings of PMV including the absence of one papillary muscle, a funnel-shaped mitral valve, a doming-shaped elongated chordae tendineae, and a pear-shaped left atrium.[18] A supramitral ring in a form of membranous or fibrous shelf is often an integral part of the PMV, thereby, significantly reducing the effective orifice area of the mitral valve.[68]

The characterized single papillary muscle which receives all chords confirms true PMV. Conversely, two papillary muscles with all chordae inserting into one muscle and the other being hypoplastic indicate a parachute-like mitral valve.

According to the literature review, the mean mitral valve annulus diameter was 8.2 (range, 7 to 10) mm, which corresponds to a z-score of -0.665 on the basis of the normal range for newborns (10±2.6 mm).[66]

**Surgical treatment and outcomes**

Serraf et al.[69] reported that PMV-related mitral stenosis often caused failure of biventricular repair in newborns with borderline small left ventricles, thus strengthening the importance of the left ventricular inflow status in decision making for either a uni- or a biventricular treatment strategy.

Balloon mitral valvuloplasty decreased the peak and the mean mitral valve gradients by a median of 33% and 38%, respectively; however, 54.5% (6/11) patients with a supravalvular mitral ring developed significant mitral regurgitation following balloon mitral valvuloplasty.[6]

Mitral repair has been a preferred procedure as opposed to MVR. In some patients, repair of a stenotic PMV was performed through a papillary muscle incision and leaflet fenestration.[70] In children, MVR shows several drawbacks, such as high operative mortality, significant incidence of complete heart block and pacemaker implantation, lack of prosthetic valves with sizes and with growth potential that are suitable for small children, difficulties in postoperative anticoagulant therapy, and rapid bioprosthetic valve deterioration.[71]
Shone et al.[1] reported that mitral valve obstruction was the most critical problem of this lesion. The severity of the mitral valve obstruction was found to be inversely correlated with long-term outcomes, and the operative mortality of patients with Shone syndrome was eventually adversely affected.[66] However, Marino et al.[89] found no significant association between progressive mitral stenosis and PMV type, dominant papillary muscle, sex, or any surgical or interventional therapies.

As PMVs are usually not isolated lesions and are characterized by a constellation of pathological changes of the mitral valve leaflets, annulus, commissures, subvalvular apparatus, and supravalvular mitral ring, the majority of patients need one or more surgical operations and the reintervention rate is high.[39]

In conclusion, about two-thirds of parachute mitral valve patients require surgical treatment of the mitral valve lesions. Parachute mitral valves are curable by mitral valve repair in most cases, and mitral valve replacement is indicated only for patients with severe mitral valve lesions.

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