

# Parachute Mitral Valve: Anatomy and Operation

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## ABSTRACT

Parachute mitral valve is a rare congenital heart defect characterised by a distorted mitral geometry with a single papillary muscle for all mitral chords to attach. It may develop in isolation or as a constellation of Shone syndrome. By comprehensive retrieval of the pertinent articles published since 2000, 22 articles with 149 cases of parachute mitral valve were recruited into this study. The present article revealed that most patients with a parachute mitral valve had a stenotic or regurgitant mitral valve, which is often associated with left heart obstructions, thereby leading to haemodynamic compromise. Therefore, multiple surgical maneuvers are warranted for such patients. Mitral valve repair is preferred over mitral valve replacement for the mitral valve abnormalities so as to avoid the associated complications of valve replacement procedure. The patients' outcomes are satisfactory with a total event-free survival of surgical patients of 84.8%.

**Key Words:** *Cardiac surgical procedures, Congenital heart defects, Mitral valve.*

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## INTRODUCTION

Parachute mitral valve (PMV) is defined as a single papillary muscle, either anterolateral or posteromedial, to which all mitral chords are attached.<sup>1</sup> The valves with only one papillary muscle are termed as true PMVs, while those with two papillary muscles with one of them being hypoplastic are defined as parachute-like mitral valves.<sup>2</sup> Both PMVs and parachute-like mitral valves have congenital anomalies of the papillary muscles.<sup>3</sup> PMV may occur in isolation or in association with other congenital heart defects.<sup>4</sup> It can be a constellation of Shone syndrome, characterised by four cardiovascular defects, including supravulvar mitral membrane, valvular mitral stenosis by a PMV, subaortic stenosis and aortic coarctation.<sup>5</sup> Walter *et al.* described that in 137 patients with congenital mitral stenosis, 48 (35%) patients were Shone syndrome.<sup>6</sup>

Left heart obstruction is very common in patients with a PMV. Ma *et al.* categorised left heart obstruction into 2-, 3-, and 4-level obstructions, *i.e.* the obstructions may occur at 2-4 levels of mitral valve, supra mitral valve, aortic valve, supra aortic valve, subaortic valve and aortic coarctation.<sup>7</sup> The coexistence of the lesions in the patients can be the cause of more complex haemodynamic changes, and thus warrants surgical corrections.<sup>7</sup>

It has been reported that the severity of the mitral valve obstruction correlates negatively with long-term outcome.<sup>7</sup> Multilevel left-heart obstructions and mitral valve disorders are significant predictive factors of poor outcomes of these patients.<sup>8</sup> The treatment of PMV and the associated disorders as well as the prognoses of the patients are insufficiently described to date.<sup>8</sup>

The aim of this article is to discuss the anatomic features of PMV and associated congenital heart defects, surgical management, and patients' outcomes.

## METHODOLOGY

This meta-analysis was conducted by following the preferred reporting items for systematic reviews and meta-analyses (PRISMA) statement guidelines. Publications published from January 2000 to August 2019 were systematically searched in the PubMed, Highwire Press, and the Cochrane Library databases. The MeSH terms and keywords used to identify articles included parachute mitral valve, supravulvar mitral ring, single papillary muscle, Shone syndrome, congenital mitral stenosis, surgical operation, mitral valve repair, and mitral valve replacement, published in English language. The screening of the bibliographic references helped the completeness of the literature retrieval. Fifty-five articles were found related to the topic and keywords in the literature search; and 22 articles, which met the inclusion criteria during preliminary assessment, were included in this review. The exclusion criteria were: articles describing the diagnosis of PMV other than surgical treatment or patients not indicated for an operation ( $n=28$ ), ambiguous patient information of operation for PMV ( $n=4$ ), and parachute-like asymmetric mitral valve ( $n=1$ ).

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The data independently extracted from each study were the study population, demographics, clinical diagnosis, mitral morphology, surgical procedures and patients' outcomes.

An IBM SPSS version 22.0 was used for statistical analysis. The measurement data were expressed in mean  $\pm$  standard deviation with range and median values; and were compared by independent sample *t*-test. The categorical variables were compared by Fisher exact test.  $P < 0.05$  was considered statistically significant.

## RESULTS

In 22 articles<sup>6-27</sup> including 7 (31.8%) original articles,<sup>6-8,10,12,17,18</sup> and 15 (68.2%) case reports with 148 patients were collected as the materials of this study. Gender of 62 patients were described: 38 (61.3%) were males and 24 (38.7%) were females ( $\chi^2=6.3$ ,  $p=0.019$ ).

The mean age of patients was  $19.2 \pm 22.9$  (range, 0-73; median, 9.1) years ( $n=24$ ). There was no significant age difference between the male and the female patients although the age of the females were higher than of the males ( $21.9 \pm 19.4$  years vs.  $42.1 \pm 28.5$  years,  $p=0.173$ ).

One hundred and twenty-eight (86.5%) patients had a non-syndromic PMV, while 20 (13.5%) patients had a PMV as a constellation of Shone syndrome (syndromic PMV) ( $\chi^2=2.7$ ,  $P=0.153$ ).<sup>7,16,22,25,27</sup> Thirteen presentations were described for 11 patients with dyspnoea/shortness of breath being the most common (Table I). Heart murmur was audible in eight patients, three patients had a systolic murmur,<sup>11,14,20</sup> two patients had a diastolic murmur,<sup>21,26</sup> and three patients had both systolic and diastolic murmurs.<sup>9,19,22</sup> By echocardiography, PMVs showed heterogeneous anatomical features of the mitral valve and subvalvular apparatus (Table II).

The PMVs were stenotic more than regurgitant. A surgical operation was required for mitral valve disorders in 68.6% (70/102) patients. The requirement for a surgical intervention did not differ between patients with mitral regurgitation and those with mitral stenosis (Table III).

In the surgical patients for mitral regurgitation, three (50%) patients required mitral valve replacement,<sup>11,12,19</sup> and 3 (50%) patients received mitral valve repair. The mitral valve repair techniques used were ring annuloplasty,<sup>12,24,26</sup> double-orifice technique,<sup>26</sup> and artificial chord reconstruction.<sup>24</sup> In the surgical patients for mitral stenosis, 10 (18.5%) patients had mitral valve replacement,<sup>10,12,15,18</sup> and 44 (81.5%) patients had mitral valve repair ( $\chi^2=42.8$ ,  $p < 0.001$ ). The requirement for mitral valve replacement or repair differ significantly between patients with mitral regurgitation and those with mitral stenosis (Table III).

The mitral valve repair techniques for mitral valve stenosis were mitral valvuloplasty ( $n=18$ ),<sup>7,18,23</sup> mitral valvotomy ( $n=13$ ),<sup>3,17</sup> mitral valve dilatation ( $n=2$ ),<sup>10</sup> mitral commissurotomy, papillary muscle splitting, posterior leaflet extension and ring annuloplasty ( $n=1$ ),<sup>2</sup> mitral commissurotomy ( $n=1$ )<sup>21</sup> and balloon mitral valvuloplasty ( $n=9$ ).<sup>18</sup> In 10 patients with mitral stenosis, the

surgical techniques were not described. Reinterventions included redo-mitral valve replacement ( $n=6$ ),<sup>3,10,17,23</sup> redo-mitral valve repair ( $n=1$ ),<sup>9</sup> and second mitral valve reintervention ( $n=1$ ).<sup>6</sup> A comparison of the associated congenital heart defects before surgical and non-surgical patients was shown in Table IV. The pressure gradient through the PMV was  $17.9 \pm 10.0/4.6 \pm 2.1$  mmHg<sup>14,17,19,21,22,27</sup> with a mean of  $12.2 \pm 5.0$  mmHg<sup>[9,16,20]</sup>.

**Table I. Clinical presentations of 11 patients.**

Presentation	n (%)
Dyspnoea / shortness of breath <sup>11,14,15,19-22,25,26</sup>	9 (69.2)
Chest discomfort <sup>20</sup>	1 (7.7)
Chest pain <sup>11</sup>	1 (7.7)
Cough <sup>11</sup>	1 (7.7)
Asymptomatic <sup>9</sup>	1 (7.7)

**Table II. Anatomic features of parachute mitral valves.**

Mitral valve	Anatomy
Leaflet	Thickened <sup>15,27</sup>
	Small <sup>13</sup>
	Dome-shaped <sup>15,27</sup>
	Pear-shaped <sup>15</sup>
	Cleft (posterior mitral leaflet) <sup>26</sup>
	Prolapse <sup>13,27</sup>
Annulus	Restricted diastolic motion <sup>15,21</sup>
	No thickening <sup>20</sup>
Commissure	Fused <sup>15</sup>
Chord	Elongated <sup>16</sup>
	Elongated and lax <sup>11</sup>
	Short bridging <sup>27</sup>
	Short and underdeveloped <sup>22</sup>
	Thickened and fused <sup>15</sup>
Papillary muscle	A single posteromedial papillary muscle <sup>15,16,21</sup>

The bicuspid aortic valve were stenotic in 4 patients<sup>10,25</sup> and regurgitant in 4 patients.<sup>12,21</sup> In the remaining 22 patients, the bicuspid aortic valves were with a normal function, otherwise the valvular function was unspecified. The surgical operations for the bicuspid aortic valves were aortic valve replacement ( $n=3$ ),<sup>12</sup> open aortic valvotomy ( $n=2$ ),<sup>10</sup> and balloon aortic valvotomy ( $n=1$ ).<sup>10</sup> Aortic valve replacement was also performed in another patient in whom the aortic valve structure was not indicated as a bicuspid or a tricuspid one.<sup>7</sup>

Other surgical operations included aortic arch repair in 13 patients,<sup>17</sup> atrial septal defect closure in 3 patients,<sup>10,17</sup> and atrial septal defect closure with division of vascular ring,<sup>10</sup> pulmonary artery banding,<sup>17</sup> supraaortic ridge excision,<sup>4</sup> unbalanced atrioventricular septal defect patch repair, atrial septal defect closure with right-sided valve annuloplasty,<sup>13</sup> patch aortoplasty for supravalvar aortic stenosis,<sup>10</sup> pulmonary arterioplasty,<sup>10</sup> and pulmonary valve replacement<sup>10</sup> in one patient each.

The patients were at a follow-up of  $176.2 \pm 160.0$  (range, 2-480; median, 192) months ( $n=15$ ). Outcomes were indicated for 92 patients: 76 (82.6%) patients recovered, 2 (2.2%) patients improved with moderate mitral regurgitation,<sup>21,22</sup> 2 (2.2%) patients were complicated with re-stenosis after coarctation,<sup>7</sup> 4 (4.3%) patients required reinterventions (with unstated cause in 2 patients<sup>12</sup> and reintervention for balloon dilation of the mitral valve in 2 patients<sup>5</sup>) and 8 (8.7%) patients died.<sup>3,17</sup>

**Table III: A comparison of the associated congenital heart defects between surgical and non-surgical patients.**

Mitral valve disorder	Total	Surgical	Non-surgical	Reintervention	2 <sup>nd</sup> Reintervention
Mitral regurgitation	9 (100)	6 (66.7)	3 (33.3)	0 (0)	0 (0)
Mitral stenosis	93 (100)	64 (68.8)	29 (31.2)	7 (10.9)	1 (1.1)
$\chi^2$	138.4	0.0	0.0	0.7	0.1
p	<0.001	1.000	1.000	1.000	1.000

**Table IV. A comparison of the associated congenital heart defects between surgical and non-surgical patients.**

Associated disorder	Total	Surgical	Non-surgical	$\chi^2$	p
Supra mitral valve ring	10 (100)	6 (60) <sup>7,9,10,16,22</sup>	4 (40) <sup>10,12,23,25</sup>	0.8	0.656
Ventricular septal defect	15 (100)	15 (100) <sup>9,10,14*,16,17</sup>	--	--	--
Pulmonary artery hypertension	4 (100) <sup>9,14,15,22</sup>	--	--	--	--
Subaortic obstruction	7 (100)	4 (57.1) <sup>10,12,20</sup>	3 (42.9) <sup>10</sup>	0.3	1.000
Bicuspid aortic valve	22 (100)	6 (27.3) <sup>10,12</sup>	16 (72.7) <sup>10,11,12,21-23,25,27</sup>	10.3	0.002
Coarctation of the aorta	34 (100)	34 (100) (surgical in 25 <sup>7,10,12,23,27</sup> , percutaneous angioplasty and/or stent placement in 9 <sup>7,11,17,20,22,26</sup> , reintervention by using balloon angioplasty in 2 <sup>10</sup> )	--	--	--
Patent ductus arteriosus	5 (100)	4 (80) <sup>10,13</sup>	1 (20) <sup>12**</sup>	3.6	0.206

\* Ventricular septal defect closed by using an Amplatzer septal occluder.<sup>14</sup> \*\*Treatment of patent ductus arteriosus was not indicated.<sup>12</sup>

The reintervention time was  $21.0 \pm 11.6$  (range, 2–40, median 24) years (n=10).<sup>12</sup> The total event-free survival was 84.8% (78/92).

## DISCUSSION

Paediatric patients with congenital mitral stenosis (most of which are due to PMV) often have severe symptoms, while very few of them are asymptomatic. Surgical intervention is indicated for the symptomatic and haemodynamically compromised patients.<sup>21</sup> The diagnosis of PMV and the associated lesions can be made by echocardiography and angiography, which may reveal severe mitral stenosis/regurgitation with a parachute deformity.<sup>28-30</sup> Most patients present with mitral stenosis of varying degrees of severity, and rarely present with severe mitral regurgitation.<sup>11</sup> This study also disclosed a prevailed stenotic mitral valve.

PMV can be misdiagnosed as mitral valve prolapse or subaortic membrane.<sup>31</sup> Therefore, careful screening by recognising the parachute configuration of the mitral valve and the solitary papillary muscle on medical imaging is mandatory.

Mitral valve replacement poses clinical and technical challenges in terms of the small-size annulus and lifetime anticoagulation; whereas, mitral valve repair is preferred owing to its advantages in preserving the mitral valve structures and left heart geometries and avoiding possible complications of mitral valve replacement.<sup>21</sup> Moreover, mitral valve repair allows undisturbed somatic and valve growth, delays the need for future valve replacement and avoids anticoagulation.<sup>32</sup> Surgical procedures of mitral valve repair can be resection of supravulvar membrane, commissurotomy, papillary muscle vertical incision, chordal fenestration, muscular resec-

tion and instrumental annular dilation.<sup>33</sup> Only papillary muscle incision procedure may sometimes be associated with recent or later mitral regurgitation.<sup>21</sup> On the contrary, mitral valve replacement often complicates postoperative anticoagulation therapy and probability future reoperations.<sup>21</sup> As it was demonstrated in this article, the characteristic anatomical changes of PMVs were more in the mitral leaflet than in other mitral valve structures, thus mitral valve repair was more popular than valve replacement.

Younger age at operation, severity of mitral disorders, and left ventricular outflow tract obstruction are proved to be independent risk factors of reoperation and mortality.<sup>34</sup> In this study, an elder patient age at operation (at a median age of 9.1) should be a contributing factor of a higher event-free survival and lower reintervention and mortality rates.

The present study revealed that PMVs were associated more with mitral stenosis than mitral regurgitation. In most cases of PMVs, a surgical intervention with either mitral valve replacement or repair was necessary, while a close follow-up was advised in PMV patients with trivial or mild mitral stenosis or regurgitation. The associated congenital heart defects, especially the left heart obstructions, such as supra mitral valve ring, subaortic obstruction, stenotic bicuspid aortic valve, and coarctation of the aorta, often warrant a surgical treatment. The total event-free survival for the surgical patients was 84.8%.

## CONCLUSION

Most patients with a PMV have a stenotic or regurgitant mitral valve, which is often associated with left heart obstructions and haemodynamic compromise, and require multiple

surgical maneuvers. Mitral valve repair is preferred over mitral valve replacement for the mitral valve abnormalities. The patients often have a promising outcome.

#### CONFLICT OF INTEREST:

Author declared no conflict of interest.

#### AUTHOR'S CONTRIBUTION:

SMY: Contributed to the conception and design of the work; acquisition, analysis, and interpretation of data for the work; drafted the work and revising it critically for important intellectual content; finally approved the version to be published; agreed to be accountable for all aspects of the work in ensuring that questions related to the accuracy and integrity of any part of the work are appropriately investigated and resolved.

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