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## Case Reports

# Parachute mitral valve and Pacman deformity of the ventricular septum in a middle-aged male



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## ABSTRACT

Parachute mitral valve and Pacman heart (incomplete muscular ventricular septal defect) are rare congenital deformities usually reported in infants and children. Very few adult patients with these anomalies are reported but the association of the two has not been described. This report describes a 56-year-old male with exertional dyspnea who was detected to have moderately severe mitral regurgitation and mitral stenosis. Typical parachute deformity of the mitral valve with a reduced opening and common attachment of all the chordae to a single posteromedial papillary muscle was evident. The chordae were elongated, lax, and redundant, which is atypical for this anomaly. Incidentally, detected aneurysm of the basal muscular interventricular septum (Pacman deformity or incomplete triangular septal defect) was also present.

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## 1. Introduction

True parachute mitral valve (PMV) is characterized by a unifocal attachment of the mitral chordae tendinae to a solitary central papillary muscle resulting in mitral inflow obstruction and/or regurgitation.<sup>1</sup> Very few adult patients with this isolated anomaly are reported.<sup>2–4</sup> Pacman heart or partial defect of the muscular inter-ventricular septum with variable size and shape during phases of cardiac cycle is also a rare anomaly, which can be congenital or occasionally acquired.<sup>5–7</sup> This report describes a 56-year old middle-aged male with this combination of rare congenital malformations. 2D transthoracic echocardiography was the main diagnostic modality

with some incremental value provided by the 3D echocardiography.

## 2. Case report

A 56-year-old, middle-aged person was incidentally detected to have a heart murmur, 20 years back, and open-heart surgery was advised. As he was asymptomatic, he preferred conservative management. Of late, he had developed exertional dyspnea and fatigue. There was no history of orthopnea or paroxysmal nocturnal dyspnea. Physical examination revealed a well-built man with supine right upper arm blood pressure of 150/90 mmHg, regular pulse rate of 78 beats/min,

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and no evidence of heart failure. Precordial examination showed normal heart sounds, a 3/6 pan-systolic apical murmur with radiation towards base, followed by a low-pitched diastolic rumble. No opening snap or third sound was audible. A 12-lead electrocardiogram revealed sinus rhythm with a heart rate of 80/min, normal atrio-ventricular conduction, and left ventricular hypertrophy with left atrial overload. The chest skiagram showed mild enlargement of cardiac silhouette and left atrial prominence. Routine biochemical tests were normal. He underwent detailed 2D and 3D transthoracic echocardiographic examination.

2D echocardiography showed enlarged left ventricle, dilated left atrium, normal ejection fraction (62%), and slightly thickened mitral leaflets. An interesting observation was an aneurysm-like incomplete defect of the basal muscular interventricular septum, which showed closure of its mouth during systole and opening during diastole like the Pacman (Fig. 1). Largest size of the defect was seen in the early diastole. There was no interventricular communication by color flow mapping.

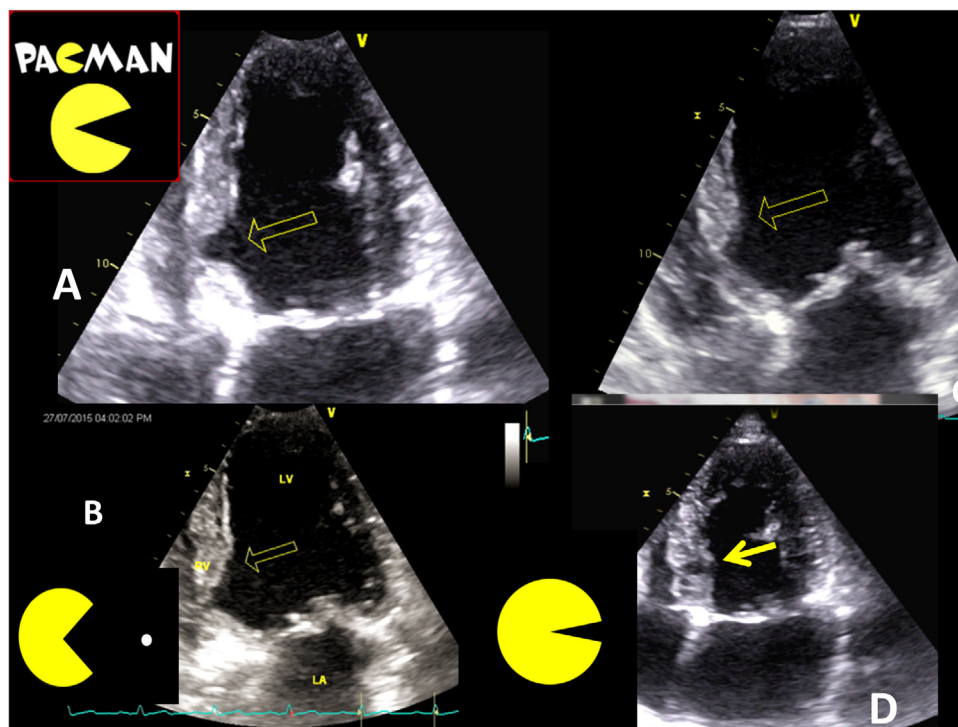
The mitral funnel was pear shaped in diastole with tunnel-like apex, eccentrically oriented with long redundant chords attached to a solitary papillary muscle originating from the inferolateral wall (Figs. 2-3). The posteromedial and solitary papillary muscle was also relatively rudimentary with a short stump and a single head. A small muscular ridge or trabecula was present at the location of anterolateral papillary muscle without any chordal attachment. 3D echocardiographic perpendicularly sliced views revealed a solitary papillary muscle and a mitral valve area of 1.7 cm<sup>2</sup>, measured using planimetry.

Color flow interrogation of the mitral orifice showed a narrow jet of the antegrade diastolic flow and an eccentric wall-hugging jet of the mitral regurgitation. Continuous-wave Doppler examination also showed dense jet of pan-systolic mitral regurgitation and a mean transmitral diastolic gradient of 7 mmHg (Fig. 4). There was trace tricuspid regurgitation.

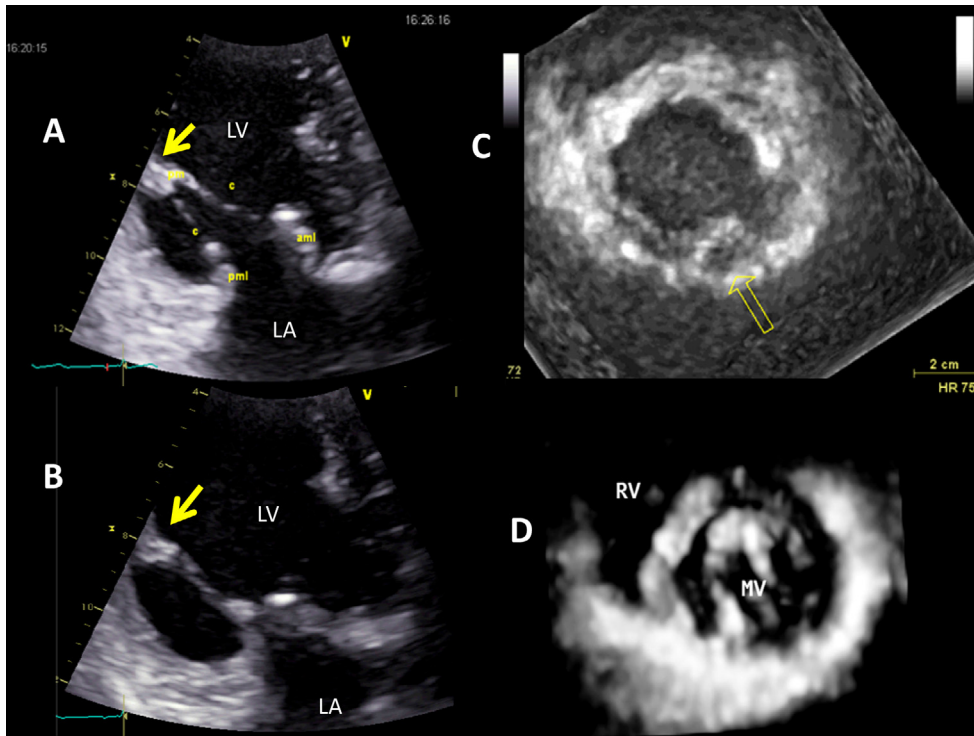
The patient has been advised mitral valve replacement.

### 3. Discussion

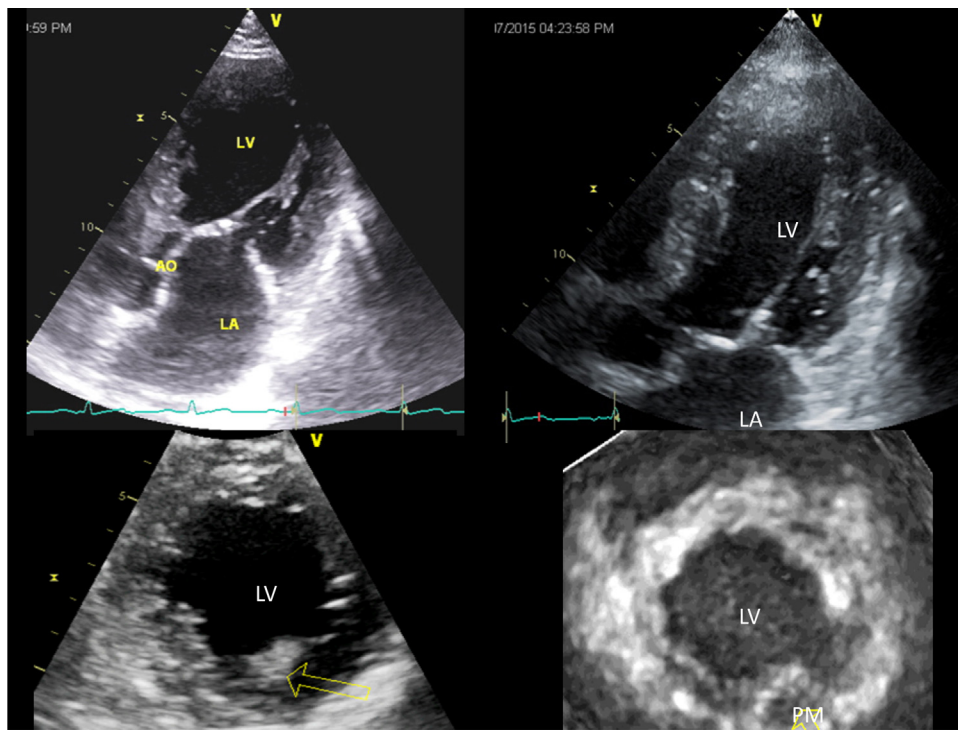
Unifocal insertion of the tensor apparatus of the atrio-ventricular valve has been labeled as the parachute deformity by Jesse Edwards and his group.<sup>1</sup> The analogy to a parachute is suggested by the shape of the deformed valve. The leaflets resemble the canopy of a parachute, the chordae, its shrouds or strings, and the papillary muscle, the harness. The pathognomonic 'pear' shape of the mitral or tricuspid valve is seen in the four-chamber view, with the atrium forming the larger base of the pear and the leaflets the apex. Dominantly but not entirely focalized insertion of chords in absence of a single muscle group has been called the parachute-like asymmetric valve, which is probably much more common than is appreciated by imaging techniques.<sup>8</sup> The anomaly was initially described as a part of the complex having other obstructive lesions of the left heart.<sup>9</sup> In three-fourth of the cases, solitary papillary muscle is a posteromedial one, situated more or less centrally. Data from the literature suggest that the valve can be distinguished on the basis of morphological features as either a parachute-like asymmetrical mitral valve or a true PMV. This occurs due to



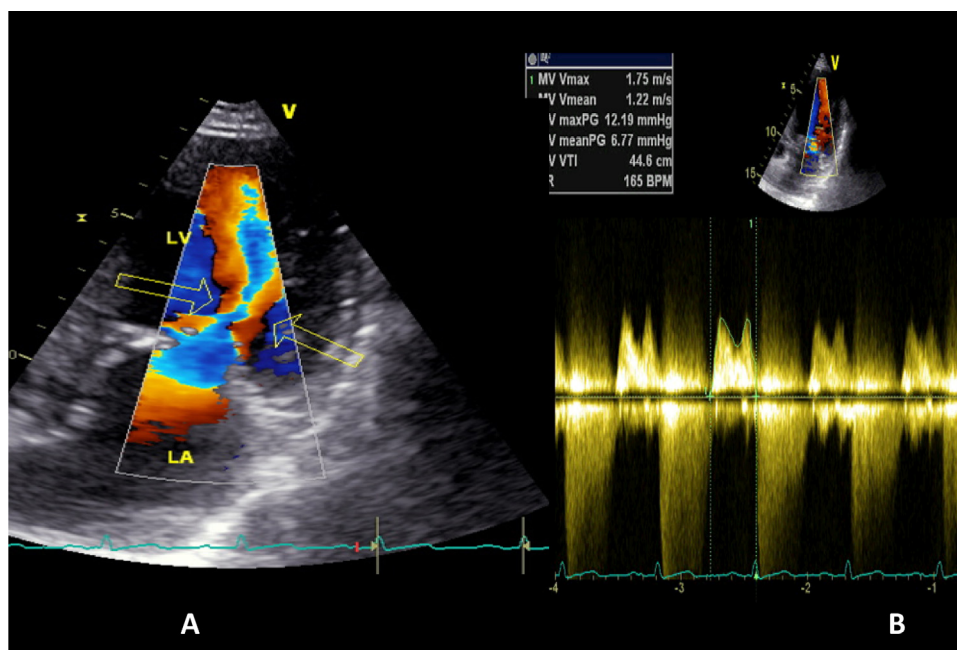
**Fig. 1** – Modified apical 4-chamber views showing partial defect of the basal muscular interventricular septum (arrows) in various phases of the cardiac cycle. In panel D, it virtually closes during end systole. The largest size is seen during early diastole (panel B). Pictures of Pacman are shown along with it.



**Fig. 2** – Transthoracic commissural views (panels A and B) showing unifocal attachment of the chordae tendinae to the posteromedial papillary muscle (solid arrows) in diastole (A) and in systole (B). A ridge of tissue without chordal attachment is seen in the middle of anterolateral wall. Panel C shows solitary papillary muscle in 3D short-axis view (blank arrow). Panel D shows parachute-like vertically placed opened mitral orifice in diastole with an orifice area of 1.7 cm<sup>2</sup> in a 3D format.



**Fig. 3** – Upper two panels show mitral valve apparatus in the apical long-axis view. Note the eccentric pear-shaped mitral orifice during diastole (left upper panel) with chords attached to the posteromedial papillary muscle. The lower two panels show a single central papillary muscle in 2D (left lower) and 3D (right lower) short-axis format.



**Fig. 4 – Transmittal color Doppler jet during diastole (A). Note the narrow jet due to narrow interchordal spaces. Panel B shows continuous-wave Doppler examination with dense spectrum of mitral regurgitation and a mean gradient of 7 mmHg.**

disturbed delamination of the anterior and posterior parts of the trabecular ridge (which normally forms anterolateral and posteromedial papillary muscles, respectively) between the 5th and 19th week of gestation, thereby forcing these embryonic predecessors of the papillary muscles to condense into a single papillary muscle.<sup>10</sup> The chordae tendinae in PMV are often underdeveloped and hence short, thick, and adherent causing decreased mobility of the valve leaflets and reducing the size of mitral orifice.<sup>2</sup> Narrowing of the interchordal spaces due to unifocal insertion results in a smaller secondary mitral orifice causing mitral inflow obstruction. Rarely the chordae tendinae may be long and lax, precluding complete coaptation of the leaflet cusps, as in our case, resulting in mitral regurgitation. Uncommonly there may be no functional abnormality of the mitral valve apparatus.

Adult patients with isolated PMV usually present with dyspnea and have hemodynamically significant lesions of variable severity across mitral valve.<sup>2</sup> However, PMV may be incidentally diagnosed during echocardiography with normal hemodynamics across the mitral valve.<sup>4</sup> Such patients generally require no medical or surgical treatment. Mitral valve surgery when feasible needs to be performed only in those patients with hemodynamically significant stenosis or regurgitation. Surgical correction of associated congenital cardiac lesion should be performed only if such lesions are hemodynamically significant and account for symptoms.

Aneurysm of the muscular interventricular septum or Pacman heart appears like an incomplete septal defect. It can be sporadic or familial and has also been described with other congenital anomalies.<sup>5,6</sup> It changes its shape during phases of cardiac cycle and may become slit-like or even absent during systole. The name is derived from an Arcade video game developed in Japan wherein the Pac-Man swallows its

perceived enemies or ghosts. Proposed pathogenesis is similar to that of a muscular septal defect including an occasional case secondary to myocardial infarction.<sup>7</sup> Complications can be in the form of conduction disturbances, rupture, and disturbed systolic function. Association with PMV has not been reported but may have some common pathogenetic mechanism involving embryonic trabecular ridge. In general, the two lesions do not affect each other in any significant way.

In summary, with better imaging techniques, rare congenital anomalies like PMV and Pacman heart are likely to be recognized more often even in patients with advanced age.

### Conflicts of interest

The authors have none to declare.

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