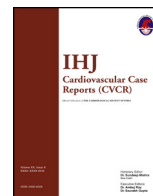




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# Parachute-like asymmetric mitral valve in an adult associated with congenital ophthalmic abnormality—A rare case diagnosed by multimodality imaging

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### ABSTRACT

A parachute mitral valve (PMV) is a congenital condition characterized by the unifocal attachment of the mitral valve chordae tendinae to papillary muscle. It is rarely found in adults. We discuss an adult case of parachute-like mitral valve that was previously diagnosed as rheumatic mitral valve disease and subsequently, using multimodality imaging, proved to be a parachute-like asymmetric mitral valve.

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## 1. Introduction

Congenital anomalies of the mitral valve are complex and often affect multiple aspects of mitral valve apparatus.<sup>1</sup> Clinically significant congenital anomalies of the mitral valve account for approximately 0.4% of all congenital heart defects.<sup>2</sup> Some of these anomalies include parachute mitral valve (PMV), anomalous mitral arcade, double orifice mitral valve etc.

PMV disease is a very rare presentation, especially in adults with very few cases reported in the literature. We are presenting a rare case of parachute-like asymmetric mitral valve which was diagnosed by Cardiac MRI and associated with some unusual ophthalmic defects.

## 2. Case report

A 30 year old female patient presented with history of breathlessness on exertion since last 4 years (NYHA class II). She was diagnosed as a case of chronic rheumatic valvular heart disease (Moderate mitral stenosis) on 2 D echocardiography and was taking treatment for the same. She had history of diminished vision for distance and near in both eyes since childhood. She was diagnosed to have bilateral fundal coloboma involving optic disc. On

examination her pulse was 78/min, blood pressure was 130/80 mm of Hg, and pallor was observed. On ophthalmic examination, she was found to have bilateral microphthalmia, microcornea and horizontal nystagmus. Her blood report was normal, except for low hemoglobin count of 8.5 g/dl. Chest X-ray and ECG were found to be normal.

2D echocardiography revealed normal mitral valve leaflets with moderate grade subvalvular obstruction with no other abnormal findings (Fig. 1A–C). The Trans oesophageal echocardiography (TEE) was suggestive of ‘single papillary muscle’ (parachute mitral valve), i.e., antero-lateral papillary muscle receiving chordae tendinae from anterior and posterior mitral leaflet (Fig. 1D–E). Cardiac MRI was performed which revealed two distinct papillary muscles in left ventricle. Postero-medial papillary muscle was non dominant receiving few chordae tendinae and antero-lateral papillary muscle was dominant formed of fusion of multiple papillary muscles and received majority of chordae tendinae. (Parachute-like asymmetrical mitral valve).

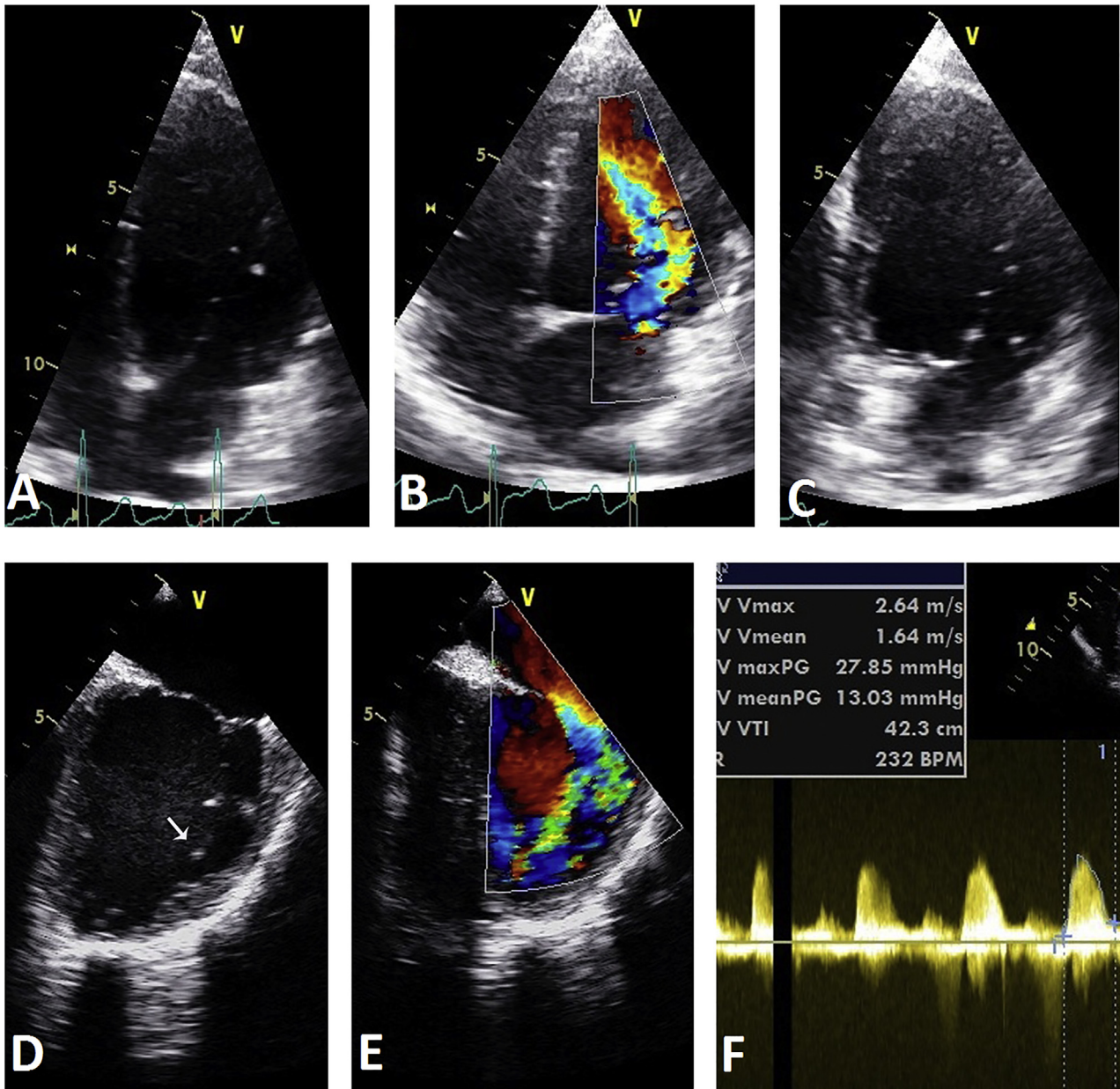
The patient was treated symptomatically with low dose beta blocker and diuretic. Her anaemia was diagnosed to be iron deficiency anaemia. She was treated with oral iron therapy. She remained asymptomatic during follow up after correction of anaemia and was treated conservatively (Fig. 2).

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**Fig. 1.** Echocardiography Images. 1A (4 chamber view), B (color Doppler across mitral valve) and C (2 chamber view) are transthoracic echocardiography images showing normal mitral valve leaflet and turbulent flow across subvalvular apparatus. 1D–F are transesophageal echocardiography images at 70° angle (mitral commissural view). 1D show single antero-lateral papillary muscle receiving all chordae tendinae. 1E and F show eccentric flow across subvalvular apparatus with significant trans-mitral valvular gradient.

### 3. Discussion

A PMV is a congenital condition characterised by the unifocal attachment of the mitral valve chordae tendinae, irrespective of the number of papillary muscles. Based on 2D Echocardiography features, the PMV complex is classified into two forms: True PMV and Parachute-like asymmetric mitral valve. The true form has a single papillary muscle that receives all chordae tendinae. The parachute-like asymmetric mitral valve has two papillary muscles, one is elongated, underdeveloped and other is well developed. Unifocal attachment of the chordate tendinae to papillary muscle could lead to either stenosis or a regurgitant lesion.

In asymmetric parachute-like mitral valves, abnormal morphologic characteristics are found in one papillary muscle. The abnormal papillary muscle is elongated, located higher in the left

ventricle, with its tip reaching to the annulus, and attached at both its base and lateral side to the left ventricular wall. This asymmetry is a persistent embryonic situation that is caused by a disturbed delamination of either the anterior or the posterior part of the trabecular ridge from the ventricular wall.<sup>3</sup>

PMV is very rare presentation in adults. It may be difficult to diagnose PMV in adults using transthoracic echocardiography. TEE can give better imaging, however it also has limitations. Cardiac MRI imaging may help in diagnosing exact mitral valve apparatus abnormality which can guide cardiac surgeon especially patients with significant symptoms requiring surgical correction.

Our patient initially diagnosed to have rheumatic mitral valve disease with moderate mitral stenosis. On subsequent transthoracic echocardiography we suspected non rheumatic abnormality in subvalvular apparatus. TEE was suggestive of true PMV. Cardiac

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