

# Accessory Mitral Valve Misdiagnosed as Vegetation: A Case Report

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## Abstract

An accessory mitral valve (AMV) is a rare anomaly of the mitral valve (MV) that often causes left ventricular outflow tract (LVOT) obstruction. We describe a young woman presenting with infrequent palpitations to our outpatient clinic. She was evaluated for mid-systolic murmur at the left sternal border. At the initial transthoracic echocardiography, vegetation on the MV was suspected. The patient was referred to our advanced echocardiography lab, where transesophageal echocardiography revealed an AMV with mild LVOT obstruction. The findings, along with extensive laboratory tests, ruled out vegetation. Additionally, she had a bicuspid aortic valve. At follow-up after 1 year, the patient was asymptomatic regarding the AMV with LVOT obstruction, and the repeat echocardiography depicted no changes compared with the previous echocardiography. Distinguishing AMVs from other MV masses, including vegetation, sometimes poses a challenge and can lead to unnecessary diagnostic and therapeutic measures. This rare MV anomaly is associated with bicuspid aortic valves.

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**Keywords:** Mitral valve; Endocarditis; Heart defects, congenital

## Introduction

Congenital abnormalities of the mitral valve (MV) include mitral stenosis, double-orifice MVs, MV parachutes, and MV clefts.<sup>1-3</sup> An accessory mitral valve (AMV) is a rare anomaly mostly resulting in left ventricular outflow tract (LVOT) obstruction.<sup>2-5</sup> We present an AMV case with mild LVOT obstruction, which was primarily misinterpreted as vegetation in an adult.

## Case Report

A 19-year-old woman presented with infrequent

palpitations to our outpatient clinic. On physical examination, the patient had mid-systolic murmur at the left sternal border. Transthoracic echocardiography (TTE) showed a mass that was attached to the anterior mitral leaflet and protruded into the LVOT, resulting in mild stenosis. For further evaluation, she was referred to our advanced echocardiography lab for transesophageal echocardiography (TEE). She had no fever, anorexia, weakness, peripheral stigmata of infective endocarditis, or a history of thromboemboli. After the first echocardiography, the patient underwent evaluations for infective endocarditis and nonbacterial thrombotic endocarditis. All the laboratory findings, including the

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erythrocyte sedimentation rate, the C-reactive protein level, the complete blood count, blood cultures, renal function tests, and screening tests for systemic lupus erythematosus, were normal.

Additionally, abdominal and pelvic ultrasonography was normal. TTE and TEE, performed at our lab, revealed a large, mobile, oval-shaped AMV ( $\approx 18$  mm in size), attached to the ventricular side of the anterior MV leaflet (Movie Clips 1 & 2). The AMV was echogenic, with an echo-free central part (cyst-like) and sharp borders that were almost oval-shaped like the MV (Figures 1 & 2).

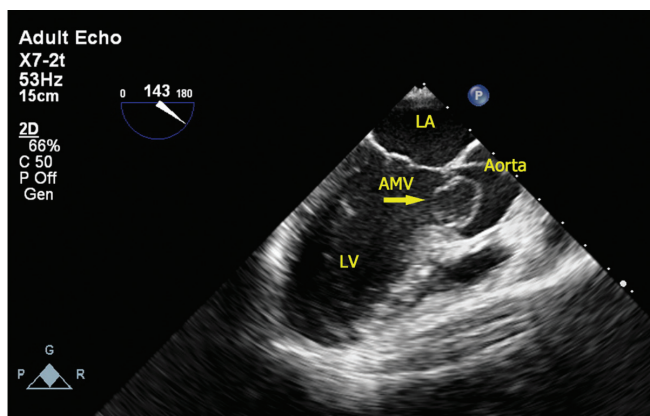


Figure 1. The image presents transesophageal echocardiography in the long-axis view. The arrow shows a large, cyst-like mass on the ventricular side of the anterior mitral valve leaflet during the systole.

LA, Left atrium; LV, Left ventricle; AMV, Accessory mitral valve

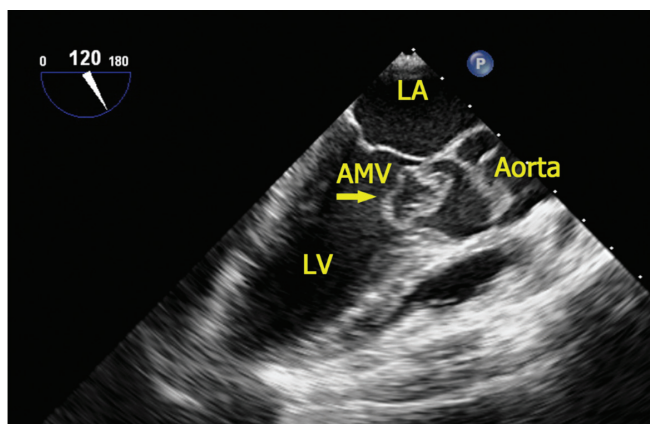


Figure 2. The image presents transesophageal echocardiography in the long-axis view. The arrow shows a large mass on the ventricular side of the anterior mitral valve leaflet during the systole. The aortic valve is bicuspid. LA, Left atrium; LV, Left ventricle; AMV, Accessory mitral valve

LVOT evaluation with color Doppler illustrated a turbulent flow with mild LVOT stenosis (mean pressure gradient=13 mmHg, peak pressure gradient=22 mmHg) (Figure 3). The aortic valve was bicuspid and dome-shaped with moderate aortic regurgitation but without stenosis. A raphe was depicted between the left and right

coronary cusps. The ascending aorta diameter was normal. Evaluations for aortic coarctation were negative, and MV function was normal and without stenosis or regurgitation. On examination, a blowing-type diastolic murmur, along with a systolic ejection sound and a mid-systolic ejection-type murmur, was auscultated at the upper left sternal border. Despite these findings, she was asymptomatic. At follow-up after 1 year, the patient was asymptomatic in terms of the AMV with LVOT obstruction, and the findings of the repeat echocardiography showed no change compared with the previous echocardiography.

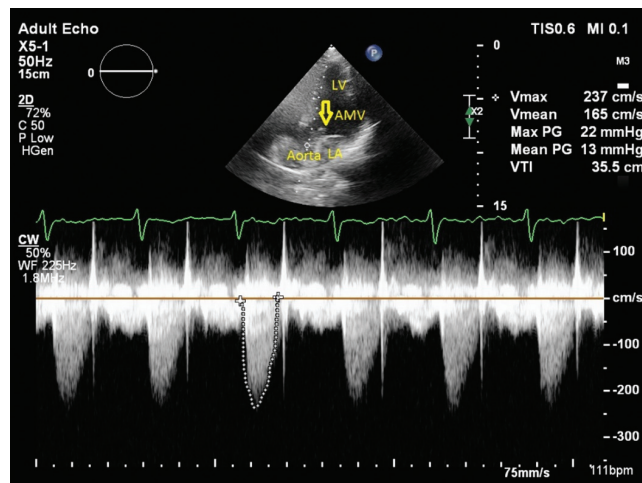


Figure 3. The image presents transthoracic echocardiography in the 5-chamber view. Continuous-wave (CW) Doppler shows a mild increase in the LVOT (peak gradient=22 mmHg). The arrow shows the LVOT mass. LV, Left ventricle; LA, Left atrium; AMV, Accessory mitral valve; LVOT, Left ventricular outflow tract

## Discussion

The differential diagnosis of MV masses includes infective endocarditis, nonbacterial thrombotic endocarditis, organized thrombi, blood cysts, and tumoral masses such as papillary fibroelastomas, myxomas, and calcified toothpaste tumors of the mitral annulus.<sup>6</sup> Some lesions have distinctive features and can be diagnosed easily. Echocardiographic findings in myxomas include narrow stalks with mixed echogenic patterns and those in papillary fibroelastomas include round heads with fine shimmering papillae. On the other hand, some masses have no specific features at echocardiography, rendering a definite diagnosis impossible, especially when the echocardiography acoustic window is suboptimal. Such uncertainties result in wide differential diagnoses and mandate extensive evaluations through laboratory and other imaging tests.

Our patient had a bicuspid aortic valve with a suspicious mobile mass, which was misinterpreted as endocarditis; she was, consequently, evaluated for both infective and



nonbacterial thrombotic endocarditis. Many congenital heart diseases have multiple different presentations and sometimes have complex lesions. These concomitant lesions must be considered in any patient with congenital heart disease to prevent unnecessary measures and costs. Most case reports have presented LVOT obstruction with significant stenosis due to AMVs, necessitating surgical resection.<sup>2-5</sup> Nonetheless, our patient had mild LVOT stenosis and was asymptomatic at 1-year follow-up, requiring no therapeutic intervention.

## Conclusion

Accessory mitral valves constitute a rare congenital heart anomaly seen as a mitral valve mass in echocardiography. Accessory mitral valves can present with or without significant left ventricular outflow tract stenosis, and they are observed mostly with other congenital anomalies such as bicuspid aortic valves.

***To watch the following videos, please refer to the relevant URLs:***

<https://jthc.tums.ac.ir/index.php/jthc/article/view/1535/1002>

Video 1. Transesophageal echocardiography shows a mass that is attached to the ventricular side of the anterior mitral valve leaflet and protrudes into the left ventricular outflow tract.

<https://jthc.tums.ac.ir/index.php/jthc/article/view/1535/1003>

Video 2. Transthoracic echocardiography shows a large mobile mass that is attached to the anterior mitral valve leaflet and protrudes into the left ventricular outflow tract.

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