

# A rare cause of the left ventricular outflow tract obstruction: accessory mitral valve tissue (RCD code: IV-1D.2o)

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## Abstract

Accessory mitral valve tissue is a rare congenital malformation, which often causes left ventricular outflow tract (LVOT) obstruction. Patients with accessory MVT and LVOT obstruction have poor exercise tolerance, fatigue, palpitations. The disease is mainly diagnosed in the 1<sup>st</sup> decade of life, when symptoms occur. Rarely diagnosis is made later, after 20–30 years of life, due to progressive course of the pathology with increasing gradient of LVOT obstruction. In cases of severe LVOT obstruction surgical management is recommended. We report a case of AMVT in young man diagnosed at the age of 31, when LVOT obstruction occurred and exercise intolerance developed. JRC D 2014; 1 (7): 13–16

**Key words:** accessory mitral valve tissue, left ventricular outflow tract obstruction

## Case report

35-year man presented with exertional dyspnoea during the last 2 months. Systolic murmur was revealed during the physical examination and aortic stenosis was suspected. ECG was unremarkable. Laboratory examination results were within normal ranges. The patient was referred to echocardiography.

Echocardiography showed that cardiac chambers dimensions and wall thickness were within normal ranges. Two-dimensional TTE revealed a free-floating mobile, sail-like accessory tissue, attached to the ventricular side of the anterior mitral valve leaflet (Figure 1). This tissue had the same echogenicity as anterior MV leaflet, and multiply chordae connected it with papillary muscle (Figure 1A). The structure moved to the left ventricular outflow tract (LVOT) in systole, systolic anterior motion of the mitral valve was present, with no evidence of pronounced interventricular septum (IVS) hypertrophy (Figure 2). Based on the echocardiographic picture, accessory mitral valve tissue (AMVT) was diagnosed.

Colour Doppler showed a turbulent flow in LVOT (Figure 3). Significant LVOT obstruction with systolic gradient of 64 mm Hg was revealed (Figure 4). Mild-to-moderate mitral regurgitation was present. No other cardiac congenital malformation was found.

Cardiac MRI was performed and proved the diagnosis (Figure 5).

Surgical removal of the AMVT was proposed, but the patient refused it. The patient is managed with beta-blocker during 18 months. Metoprolol succinatis (Betaloc) was prescribed. Dosage of 50 mg of metoprolol had not a significant effect on LVOT gradient, it remained above 60 mm Hg. Dosage of 100 mg of metoprolol had a moderate effect on the degree of LVOT obstruction, the maximal gradient decreased to 55 mm Hg, mean gradient – to 20 mm Hg (Figure. 6).

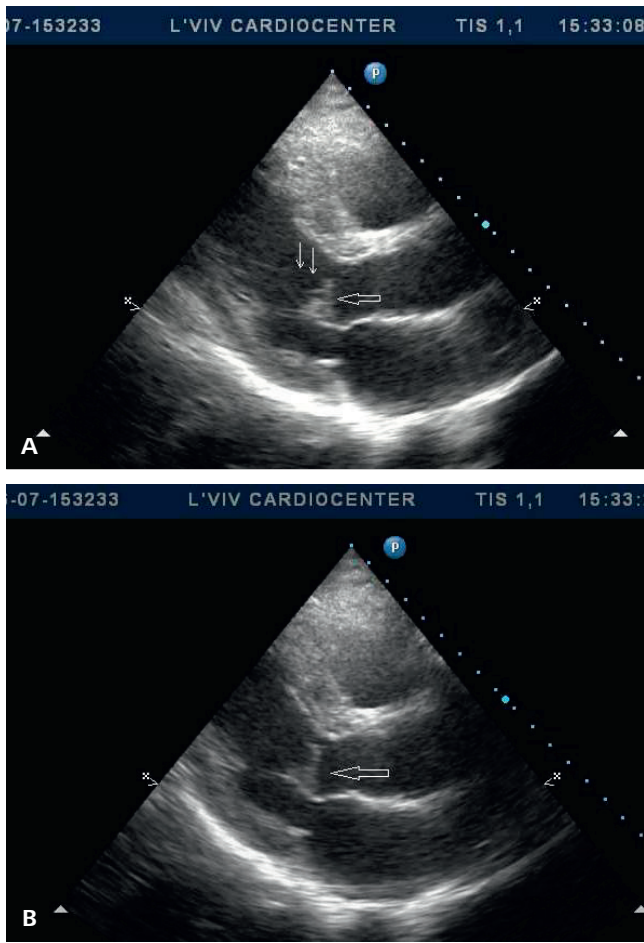
Taking into consideration the potential harmful impact of the high LV intracavitary pressure on cardiac rhythm, 24-hours ECG monitoring was performed. It showed absence of significant rhythm disorders. Only 138 premature supraventricular beats were recorded, no episode of non-sustained or sustained VT was registered.

The patient is under close follow-up, he has not complaints. The patient tolerates well his everyday activity and refuses surgical management. Antibiotic prophylaxis for infective endocarditis is recommended.

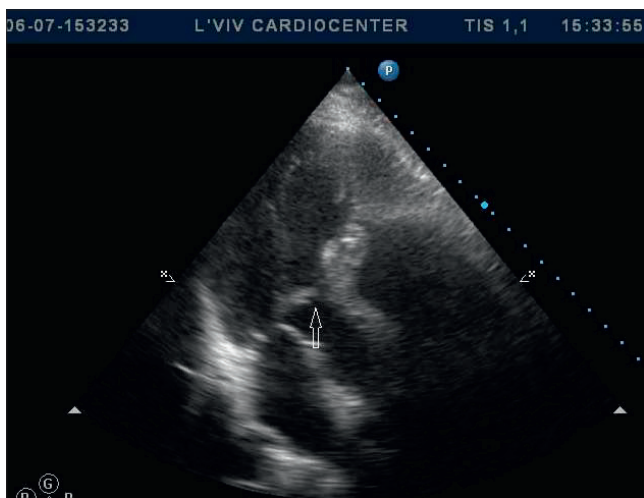
Conflict of interest: none declared.

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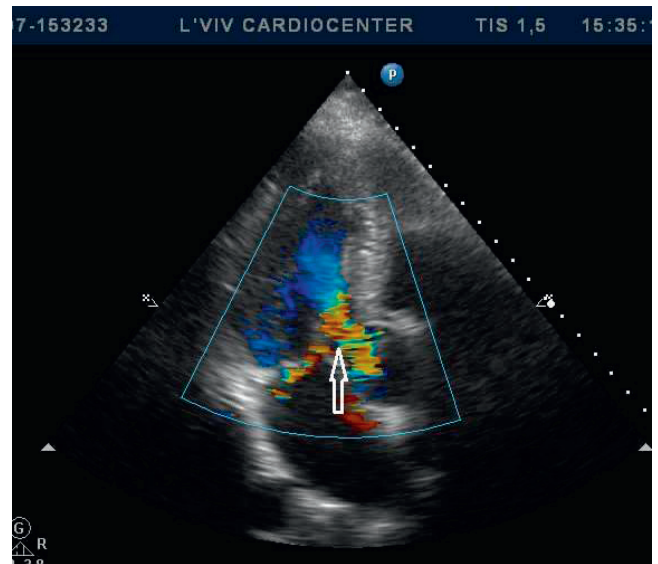
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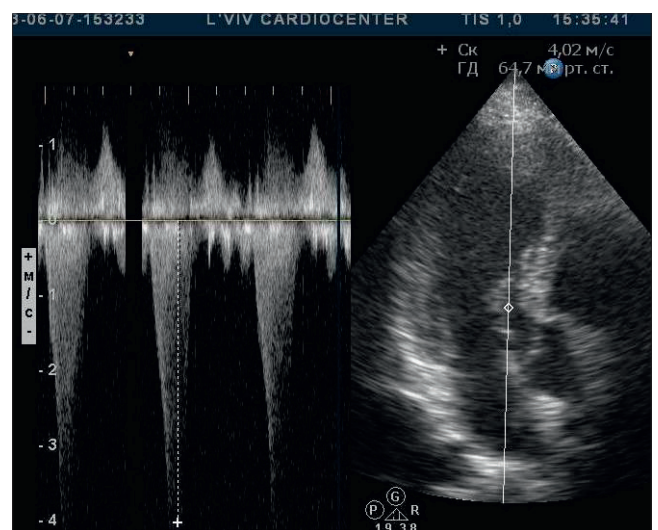
**Figure 1.** Transthoracic echocardiography. Parasternal long axis view. **A.** Accessory mitral valve tissue (thick arrow), multiply chordae (two arrows). **B.** A sail-like accessory tissue, attached to the ventricular side of the anterior mitral valve leaflet (arrow)



**Figure 2.** Transthoracic echocardiography. Apical five-chamber view. Accessory mitral valve tissue in LVOT during systole (arrow)

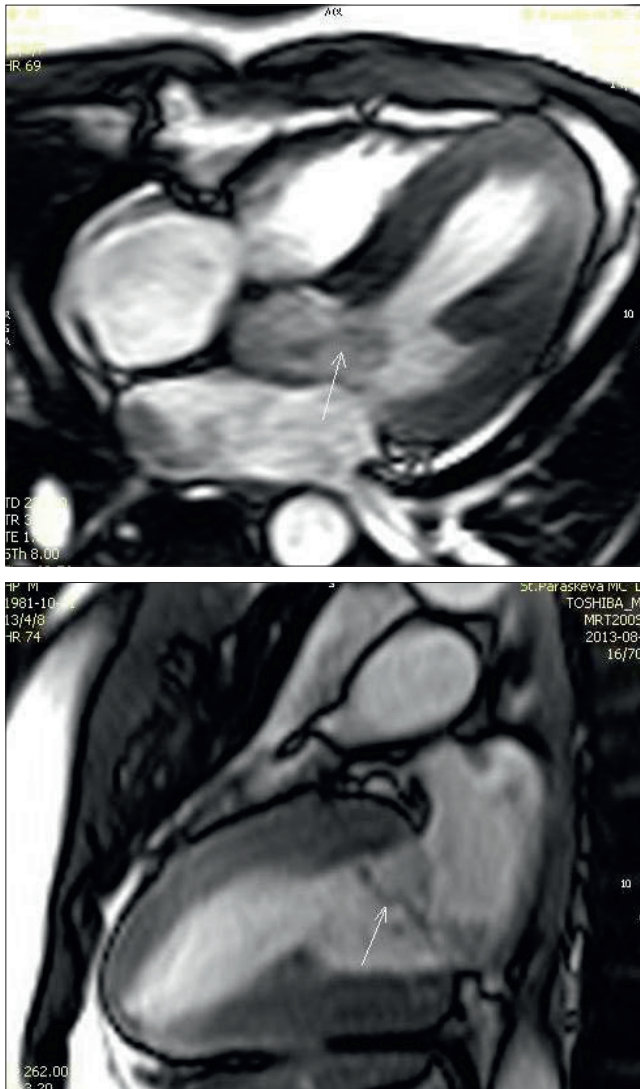


**Figure 3.** Transthoracic echocardiography. Apical five-chamber view. Colour-Doppler shows turbulent flow in LVOT, suggesting LVOT (arrow)



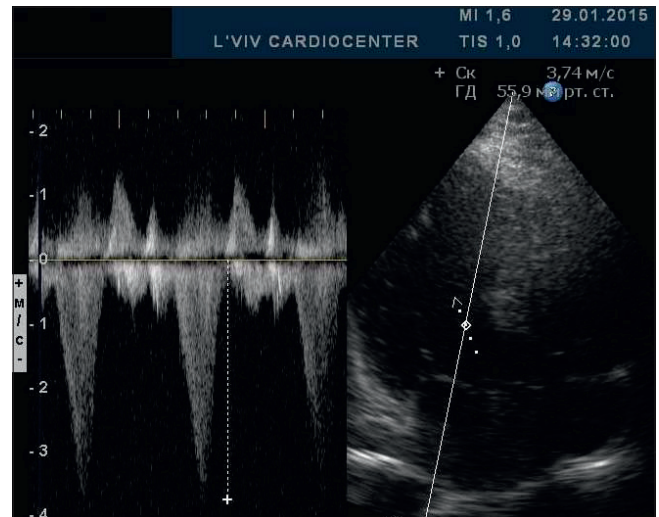
**Figure 4.** Transthoracic echocardiography. Apical five-chamber view. Continuous-wave Doppler. Late-peaking systolic gradient in LVOT, peak gradient 64 mm Hg

AMVT is a rare congenital malformation often leading to LVOT obstruction. It was first described in 1842 by Chevers, and the first operation of AMVT surgical removal has been performed in 1963 by MacLean [2, 4]. This entity can present as isolated pathology or in an association with other congenital heart diseases (bicuspid aortic valve, ventricular septal defect, transposition of great arteries, and others). It is suggested that formation of AMVT is caused by abnormal development of endocardial cushion tissue. Most of cases have been reported in children, presented with systolic heart murmur, fatigue, and exercise intolerance.



**Figure 5.** Cardiac magnetic resonance. Accessory mitral valve tissue obstructing LVOT (arrow)

The most precise review of the entity was made by Prifity and colleagues in 2003 [7]. They analyzed 90 previously reported cases. There was a slight predominance of males (51 men and 39 women), most of the patients diagnosed during the 1<sup>st</sup> decade of life. Most cases of AMVT presented with LVOT obstruction, with systolic gradient exceeding 50 mm Hg (80% of all reported patients with AMVT). Accessory MV tissue had different anatomic presentations: sac-like, parachute-like, balloon-like, leaflet-like, sail-shaped, pedunculated, membrane-like. Prifity and colleagues classified all these forms into two types: type I – fixed AMVT, and type II – mobile AMVT. Type I can have a nodular or membranous form (type IA and type IB). Type II (mobile AMVT) is classified as pedunculated (type IIA) or leaflet-like (type IIB) form. Type IIB (leaflet-like form) is divided into subtypes with rudimentary chordae tendineae or well-developed chordae tendineae (Type IIB1 and type IIB2) [7]. The most frequent form is type IIB2, often causing LVOT obstruction (27 patients; 46,5% of all presented cases).



**Figure 6.** Transthoracic echocardiography. Apical five-chamber view. Continuous-wave Doppler: systolic gradient in LVOT after therapy with metoprolol: peak gradient 55 mm Hg

In our patient the AMVT had a sail-like appearance, most similar to type IIB2, with well developed chordae tendineae, inserted into papillary muscle.

Prognosis of AMVT to the great extent depends on the presence of LVOT obstruction, which can occur in the 1<sup>st</sup> decade of life due to mass effect of the accessory tissue. LVOT obstruction can develop later due to continued deposition of fibrous tissue in the basal portion of IVS as a result of continuing turbulence produced by AMVT. In our patient the LVOT obstruction was revealed at the age of 35, thus we can suspect, that it progressed gradually [3, 6, 7, 9].

In review reported by Mangarano in 2013, LVOT obstruction occurred in 86,6% of 104 patients, in whom information about obstruction was available [5]. Symptoms of LVOT obstruction occur when the gradient exceeds 50 mm Hg. The symptoms include exercise intolerance, fatigue, syncope, heart failure, chest pain. Patients without LVOT obstruction are usually asymptomatic. The reported patient was symptomatic (fatigue, exercise intolerance) several month before diagnosis was established.

AMVT can be a potential source of thromboembolism, because highly mobile accessory tissue can accumulate platelets with further embolism. The probable association between AMVT and cardioembolic events exist [1, 10, 11], but the evidence of causative role of AMVT is not proved.

The potential influence of AMVT on cardiac rhythm exists according to the elevated LV intracavitary pressure in patients with LVOT obstruction. In literature we met only one case of nonsustained ventricular tachycardia in the patient with AMVT [5]. But according to the current guidelines on hypertrophic cardiomyopathy we should perform 24-hours ECG monitoring in patients with LVOT obstruction. So we performed it to our patient, and haven't detected any episode of VT.

Echocardiography is the best diagnostic tool for detection of AMVT and assessment of LVOT obstruction. Two-dimensional echo provides anatomic characteristics of the accessory tissue and

LV wall thickness, while Doppler-echo allows to assess LVOT gradient and to provide follow-up of LVOT obstruction.

AMVT should be differentiated with other masses which can occur in LVOT or on the anterior MV leaflet: cardiac tumors (papillary fibroelastoma or mixoma) and vegetations. Location of the mass is important: vegetations are located on the atrial side of mitral leaflet, AMVT – on the ventricular side. The fact, that AMVT usually is connected with papillary muscle or LV wall by chordal tissue, can be used for differentiation with tumors. Echocardiography is important in differentiating LVOT obstruction due to AMVT and other causes of LVOT pressure gradient (hypertrophic cardiomyopathy, discrete membranous or tunnel subaortic stenosis) [5]. Transthoracic echocardiography is usually sufficient for establishing diagnosis of AMVT, in complicated cases transesophageal examination is needed [5, 6, 8]. Cardiac MRI and CT can provide additional information about tissue characterization [12]. We performed cardiac MRI in our patient to assess all anatomical features precisely, and MRI confirmed the diagnosis (Fig 5). To our knowledge, it is only third reported case of MRI application in diagnosis of AMVT.

Several authors advocated surgical removal of AMVT in cases of LVOT obstruction, but this approach is not proved enough [3, 4]. Other authors considered that it is reasonable to treat patients medically with frequent assessment of LVOT gradient. Surgical treatment is indicated in cases of AMVT with high resting LVOT which doesn't decrease in response to medical treatment with beta-blockers and in patients undergoing correction of other congenital cardiac malformations. Surgical option is mandatory in patients with mean gradient of LVOT obstruction above 25 mm Hg. Surgical mortality is 8,9% in reported cases and is mainly caused by coexisting congenital heart disorders [7].

In our patient the resting LVOT gradient was 67 mm Hg before treatment and 55 mm Hg after treatment with 100 mg of metoprolol daily. We are performing follow-up echo every 6 month. The question about anticoagulation exists, and there is no definite answer in literature, because there is no evidence of causative influence of AMVT on embolic events in reported patients [10,11].

For all patients who met criteria of AMVT close follow-up by echocardiography is recommended for reassessment of LVOT obstruction and appropriate timing of surgery.

## Conclusion

Two-dimensional echocardiography is the main imaging modality for the morphological study of AMVT, Doppler-echocardiography is a tool for the assessment of LVOT obstruction. Cardiac MRI could be used to prove the diagnosis. AMVT should be differentiated from other causes of LVOT obstruction. LVOT pressure gradient should be followed up precisely, in cases with mean gradient above 25 mm Hg, surgical removal of AMVT is recommended.

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