Mitral annulus disjunction and Barlow's disease in a young male patient

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Introduction: Mitral annulus disjunction (MAD) is an abnormal insertion of the mitral annulus flexion line into the atrial wall, often described in association with Barlow's syndrome, a myxomatous mitral valve degeneration, causing mitral valve prolapse (MVP). It is the most common cause of primary mitral regurgitation (MR)^{1,2}. Both have been linked to increased arrhythmic risk and sudden cardiac death³.

Case report: 28-year-old patient with no previous medical history presented with palpitations, occasional shortness of breath and near loss of consciousness, especially during sport activities. Auscultatory, heart sounds were rhythmic with no murmurs except mid-systolic click. All blood tests were completely normal. Resting 12-lead electrocardiogram revealed sinus rhythm without T wave inversion or ventricular ectopic beats. Transthoracic echocardiography and transesophageal echocardiography verified borderline size and normal function of left ventricle, clear signs of MAD (Figure 1) and myxomatous changes of both anterior and posterior mitral valve leaflets with mild mitral regurgitation (Figure 2). Cardiac magnetic resonance imaging (CMR) confirmed MAD (Figure 3). The 24-hours Holter ECG monitoring did not confirm the presence of malignant ventricular arrhythmia. However, we decided to implant loop recorder for prolong rhythm monitoring of patient with MAD and Barlow's disease.



FIGURE 1. Transthoracic echocardiography. Four chamber view: green arrows are pointing at mitral annulus disjunction.

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FIGURE 2. Transesophageal echocardiography; left image 2D echocardiography and right image 3D echocardiography. Myxomatous mitral valve changes with mitral valve prolapse.

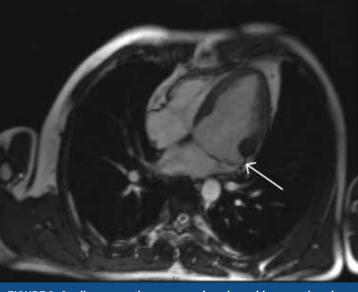


FIGURE 3. Cardiac magnetic resonance imaging: white arrow is pointing at mitral annulus disjunction.

Conclusion: Echocardiography is unavoidable method in diagnosis of mitral annulus disjunction. MAD is common finding in myxomatous valve disease and can cause symptoms of heart failure and potential life threatening ventricular arrhythmias highlighting the importance of its recognizing and treating.

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