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Echocardiography imaging of mitral annulus disjunction in a young female patient after sudden cardiac arrest

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Short title: MAD in a patient after sudden cardiac arrest

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A 38-year-old female patient was admitted to the hospital after successfully resuscitated sudden cardiac arrest (SCA). The primary registered rhythm was ventricular fibrillation. In 2006 the patient had SCA in the mechanism of ventricular fibrillation, probably in the course of myocarditis. As part of secondary prevention, a cardioverter-defibrillator (ICD) was implanted. In echocardiography exam only small mitral regurgitation (MR) was diagnosed. Unfortunately, during hospitalization at another hospital in 2020 the device was removed leaving the ventricular electrode in the tissues due to its dysfunction and the lack of system intervention since 2006.

During current hospitalization transthoracic echocardiography showed a normal left ventricular systolic function, and a severe MR due to bileaflet prolapse. Moreover, mitral annulus disjunction (MAD) measured from the left atrial wall-mitral valve leaflet junction to the top of the left ventricular wall during end-systole in the parasternal long-axis view was revealed. The distance

was 10 mm (Figure 1A; Supplementary material, *Video S1*). We did not observe an abnormal movement of myocardium (curling) but high velocity of systolic movement of mitral annulus (Pickelhaube sign) was present.

In transesophageal echocardiography severe multijet MR with pulmonary venous flow reversal was diagnosed (Figure 1B). In 2D exam bileaflet mitral prolapse (defined as superior displacement ≥ 2 mm of any part of the mitral leaflet beyond the mitral annulus) was observed (Figure 1C). 3D transesophageal echocardiography allowed to add exact information about the morphology and mobility of the leaflets. Valve leaflets thickening and a diffuse, redundant leaflet tissue with multiple segments bileaflet prolapse, chordae elongation and annular dilatation was observed (Figure 1D; Supplementary material, *Video S2*). Finally, Barlow disease with MAD was diagnosed.

Barlow's disease is more commonly observed in young and otherwise healthy patients and can be completely asymptomatic at the time of presentation [1]. MAD is associated with increased risk of ventricular arrhythmias when it is present at the posterior wall [2]. Other markers of ventricular arrhythmia are female sex, younger age, previous syncope, more premature ventricular contractions, papillary muscle fibrosis, MAD > 8.5 mm and ECG changes [3–5]. The patient hadn't have any ICD intervention from 2006 to 2020 but in our opinion the worsening of MR was the factor leading to the second SCA. The decision about implantation of subcutaneous ICD at first was made together with the cardiac surgeon who planned to operate the patient. The patient was not qualified for emergency surgery. On the other hand, it was concluded that in secondary prevention, she should receive ICD before discharge. The patient was qualified for mitral valve repair by thoracoscopy. Subcutaneous ICD implantation is not contraindication to this type of surgical access.

The patient had the subcutaneous ICD implanted and was referred to cardiosurgery department to perform surgical intervention.

Key message resulting from this case is that detailed echo examination is crucial before the decision to remove implantable cardiac devices. In this particular case, performing an echocardiographic examination before removing the device would cause replacing the device instead of removing it and could avoid re-resuscitation.

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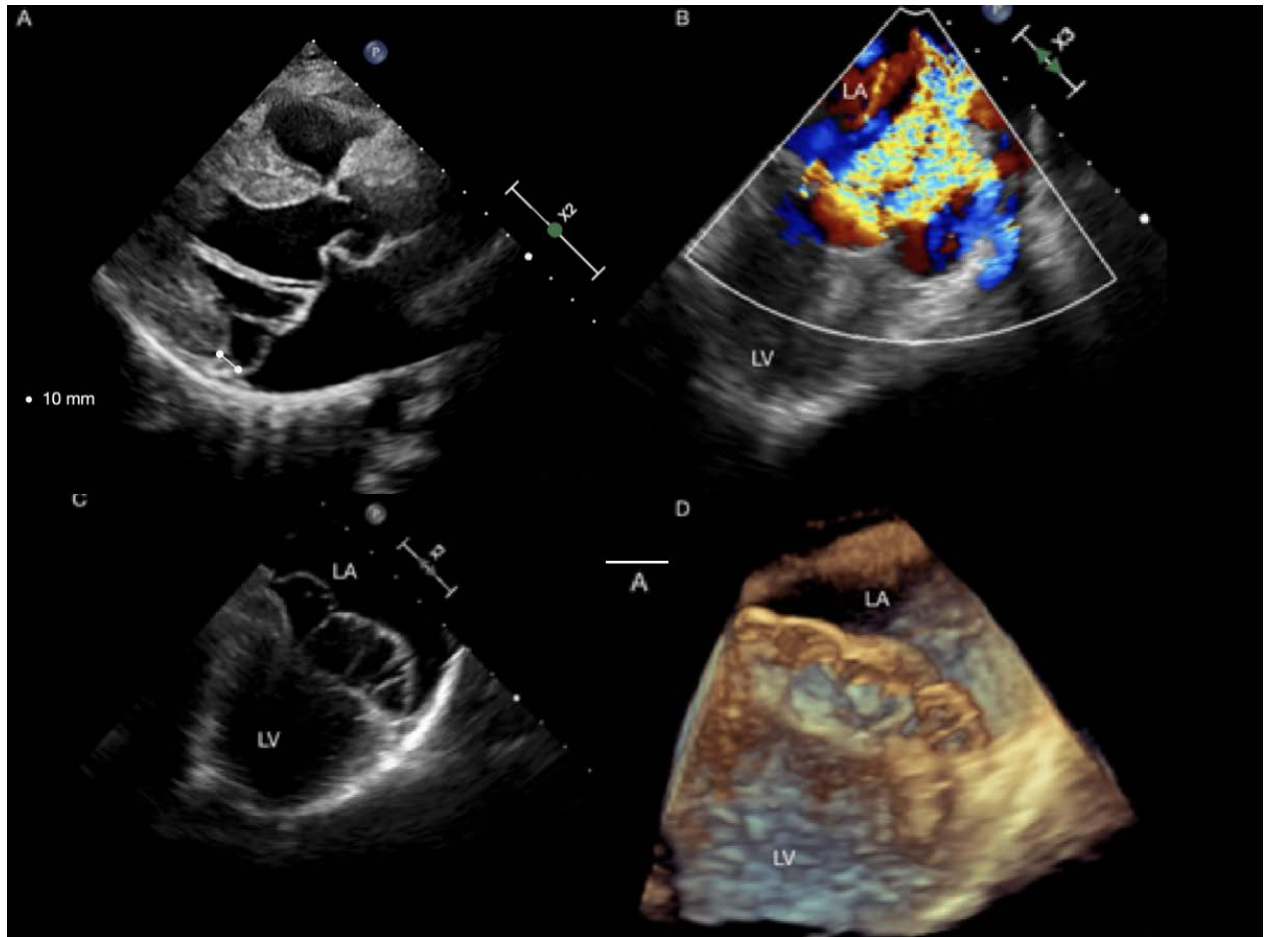


Figure 1. **A.** 2D transthoracic echocardiography, long-axis view. **B.** 2D TEE, color doppler on mitral valve. **C.** 2D TEE, two chamber view. **D.** 3D TEE, two chamber view.

Abbreviations: LA, left atrium; LV, left ventricle; TEE, transesophageal echocardiography