

ECG, an essential tool to guide arrhythmic mitral valve prolapse diagnosis

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A 35-year-old man was referred for cardiac check-up because of suspected reflex syncope. He underwent a tilt-test challenge without syncope provocation. The electrocardiography (ECG) showed sinus rhythm with polymorphic premature ventricular contractions (PVC), with right bundle branch block morphology (Figure 1). There were T-wave inversions with terminal notch of the QRS in inferior leads. An echocardiogram was performed and it showed bileaflet mitral valve

prolapse with moderate regurgitation. Left atrium was mildly dilated and left ventricular ejection fraction was preserved. Afterwards, the patient underwent prolonged Holter monitoring with frequent polymorphic PVCs (7%), and three non-sustained fast (>180 beats per minute) runs of monomorphic ventricular tachycardia. Cardiac magnetic resonance was performed showing mitral annular disjunction, and late gadolinium enhancement in posteromedial papillary muscle, and

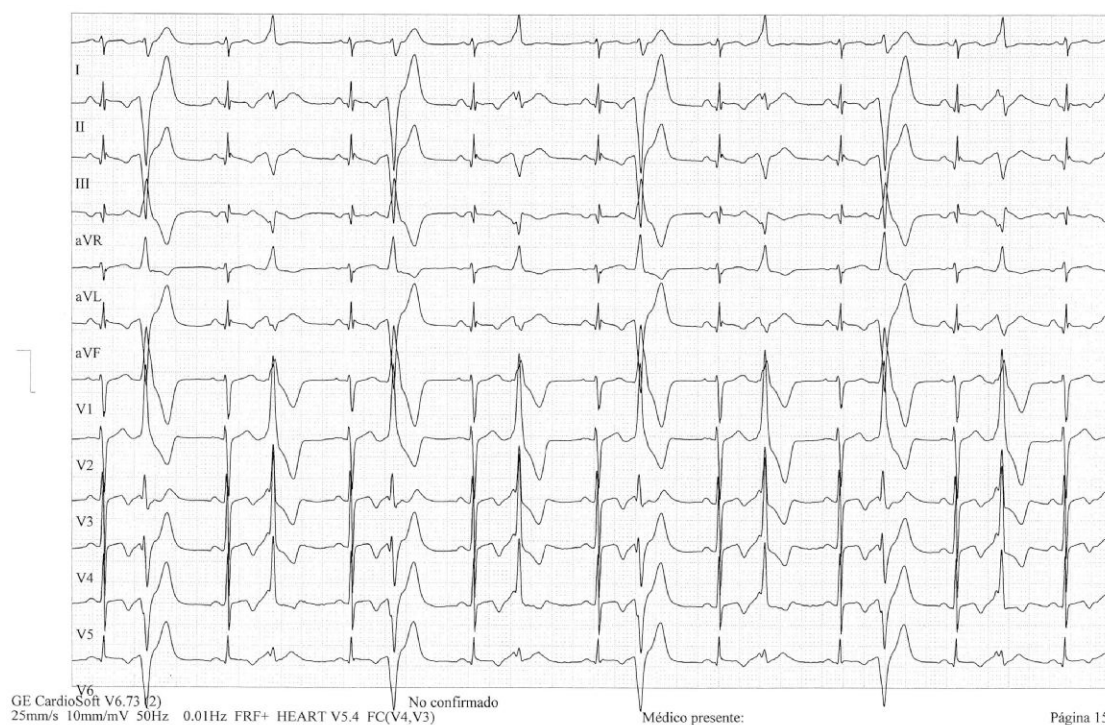


Figure 1. Baseline electrocardiogram.

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all over mitral annulus. Diagnosis of arrhythmic mitral valve prolapse was done. Implantable cardioverter defibrillator (ICD) was proposed because high-risk non-sustained ventricular tachycardia (NSVT) finding in Holter, but it was rejected by the patient. The patient accepted implantable loop recorder (ILR) implantation, and began treatment with low-dose beta-blockers, with future uptitration depending on tolerance. ICD would be considered depending on the ILR findings. Electrocardiogram is an essential tool which can suggest the diagnosis even in the absence of cardiac imaging. ECG is might!

Multiple-choice questions

1. The site of origin of the two different PVC seen in the ECG are most consistent with:
 - a. Anterior mitral annulus and posteromedial papillary muscle.
 - b. Posterior mitral annulus and left ventricular apical wall.
 - c. **Anterolateral papillary muscle and posterior mitral annulus.**
 - d. Anterolateral papillary muscle and posteromedial papillary muscle.
 - e. Posterior mitral annulus and posteromedial papillary muscle.

Answer: The first PVC corresponding to the second beat in the ECG shows right bundle branch block morphology, with rS transition in V3, and a rightward axis, suggestive of site of origin in anterolateral papillary muscle. The second PVC (corresponding for instance to the fourth beat of the ECG) shows right bundle branch block morphology with positive concordance in precordial leads, and leftward axis, suggesting a posterior mitral annulus site of origin.

2. Which one of the following is not required for the diagnosis of arrhythmic mitral valve prolapse?
 - a. **Mitral annular disjunction.**
 - b. >5% of total PVC burden.
 - c. The absence of any other well-defined arrhythmic substrate.
 - d. Complex ventricular arrhythmia.
 - e. Mitral valve prolapse.

Answer: According to the European Heart Rhythm Association (EHRA) expert consensus on arrhythmic mitral valve prolapse¹, the diagnosis criteria are the presence of mitral valve prolapse, frequent burden of PVC (>5%) in Holter monitoring, the presence of complex arrhythmia like NSVT, sustained ventricular tachycardia, or ventricular fibrillation triggered by PVC. The absence of any other well-defined arrhythmic substrate is also necessary. However, the presence of mitral annular disjunction² is not a main criterion for the diagnosis of this entity.

3. According to EHRA expert consensus on arrhythmic mitral valve prolapse, which one of the following sentences is not correct?
 - a. Cardiac magnetic resonance (CMR) should be performed in all MVP patients with a history of unexplained syncope.

- b. ILR may be advised in patients with mitral valve prolapse (MVP) and unexplained syncope in whom non-invasive ECG monitoring was not revealing or inconclusive.
- c. All MVP patients with high-risk ventricular tachycardia (VT) episodes in Holter monitoring should be considered for ICD implantation.
- d. **Reduced left ventricular ejection fraction is the most important echocardiographic predictor of arrhythmias. The progression of mitral valve disease has no role in predicting sudden cardiac death.**
- e. In MVP patients, ablation of PVCs is reasonable if triggering VF, particularly if not controlled by medications.

Answer: According to the last expert consensus on arrhythmic mitral valve prolapse, CMR must be performed in all patients that have suffered unexplained syncope, and implantable loop recorder is advised when prolonged ECG monitoring is not revealing. If there are findings of high-risk VT features, patients should be considered for ICD primary prevention implantation. If there is VF reproducibly triggered by a PVC, the patient should be referred to PVC ablation.³ Although, reduced LVEF is predictor of complex arrhythmias in this entity, the progression of mitral valve disease has a key role in increasing sudden cardiac death risk.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports*.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

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