

# Disappearance of electrocardiographic abnormalities associated with the arrhythmic pattern of a Barlow disease after surgical mitral valve repair

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## Abstract

We describe the case of a 46-year old female with a Barlow's disease (MVP) characterized by systolic curling of posterior left ventricular (LV) wall + significant mitral annular disjunction + complex ventricular arrhythmias + syncope + inverted T waves in inferolateral leads in whom a successful surgical mitral valve repair determined the disappearance not only of the echocardiographic but also the electrocardiographic abnormalities (in particular the inferolateral T waves inversion on basal electrocardiogram and the complex basal arrhythmic pattern). This case demonstrates that electrocardiographic abnormalities may disappear after the surgical correction of the mechanical stretch imposed on the inferior LV free wall by the prolapsing mitral valve leaflets. Electrocardiographic changes remain an important and easy marker to recognize for the identification of a high-risk subgroup of MVP patients.

## Case Report

A 46-year old female with a Barlow's disease (MVP)<sup>1</sup> diagnosed at the age of 20 went to a surgical mitral valve repair (annuloplasty with Cosgrove 32, neochord implantation) due to the development of left ventricular systolic dysfunction (video 1) associated with complex and symptomatic left ventricular premature beats (Figure 1A) and non-sustained runs of ventricular tachycardia (Figure 1B) and the occurrence of recurrent episodes of pre-syncope.

Mitral valve disease of this patient was characterized by thickened mitral valve leaflets, bi-leaflet mitral valve prolapse, eccentric mitral valve regurgitation (occurring mainly in meso-tele systole) and systolic curling movement of the basal portion

of the infero-posterior left ventricular myocardial wall (video 1) and a mitral annular disjunction of about 9 mm.<sup>2,3</sup>

This echocardiographic pattern was associated with the presence of negative T-waves in ECG leads D2D3aVF, V3 to V6 and premature ventricular beats with a right bundle branch block + left anterior axis deviation indicative of their origin from the posterior region of the left ventricle (Figure 2A). Coronary angiography performed before cardiac surgery revealed normal coronary arteries. A previous cardiac MRI study performed a few years before cardiac surgery to evaluate cardiac function showed focal late enhancement in the sub-endocardial layer of the infero-posterior region at the passage from basal to mid segment. At 3 months after surgery, we observed a normalization of LV function (LV ejection fraction increased from a pre-operative value of 45% to 60% post operation), the disappearance of the abnormal inward systolic movement of the basal inferior LV free wall (video 2), the absence of complex ventricular beats on the rest ECG along with the normalization of ventricular repolarization (previously negative T waves returned to a normal positive pattern (Figure 2B).

## Discussion and Conclusions

As recently proposed by Basso *et al.*<sup>2</sup> this pattern of mitral valve disease assume a form of a *so called* malignant or arrhythmic MVP<sup>3,4</sup> possibly associated with sudden cardiac death with a high prevalence of cases in young females. Uncertainty exist regarding the substrate of the electric instability in this form of MVP, even if a replacement-type fibrosis in infero-posterior myocardial region secondary to mechanical stretch of the myocardium by the prolapsing leaflets has been hypothesized as the trigger of ventricular arrhythmias.<sup>2</sup> In this patients with arrhythmic MVP we were able to demonstrate the disappearance of ECG repolarization abnormalities in particular those evident on infero/lateral leads after successful repair of the MVP associated with the normalization of inferobasal LV free wall systolic movement and the disappearance of complex ventricular arrhythmias with right bundle branch block (RBBB) + left anterior axis deviation morphology. The absence of complex ventricular arrhythmias was confirmed by a 24-h Holter monitoring performed during the 3-months follow-up period after surgery. The significance of T-wave inversion in MVP is not clear: it could be due to regional

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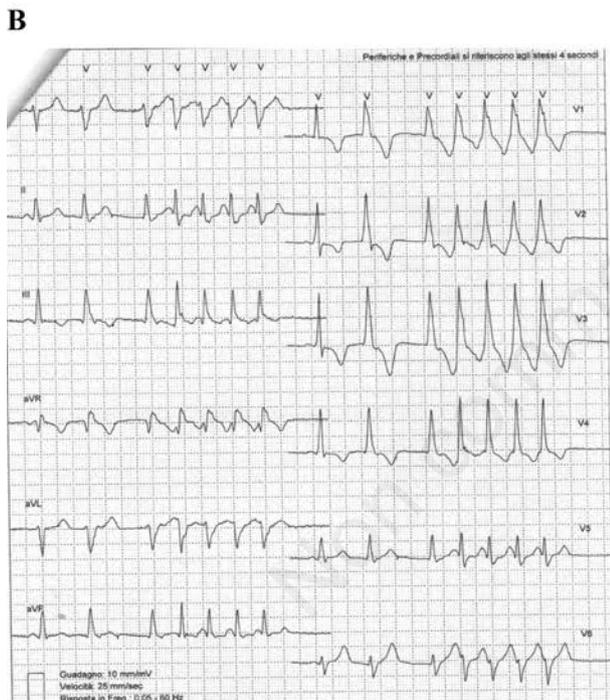
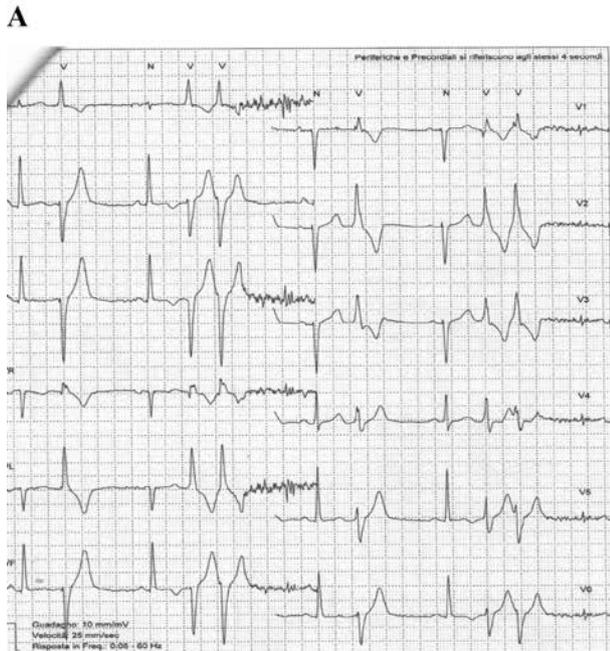
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myocardial ischemia not involving epicardial coronary arteries (that were found to be normal on pre-operative coronary angiography) or depolarization delay due to mechanical distortion of myocardial region during the occurrence of prolapse (particularly the inferior basal segment). However this case demonstrates that ECG abnormalities may disappear after the surgical correction of the mechanical stretch imposed on the inferior LV free wall by the prolapsing mitral valve leaflets, thus suggesting their non-ischemic origin. Whatever is the origin, electrocardiographic changes remain an important and easy marker to recognize for the identification of a high-risk subgroup of MVP patients (Barlow's disease+ECG abnormalities+complex ventricular arrhythmias+ history of presyncope or syncope) as recently suggested by Basso *et al.*<sup>2</sup>

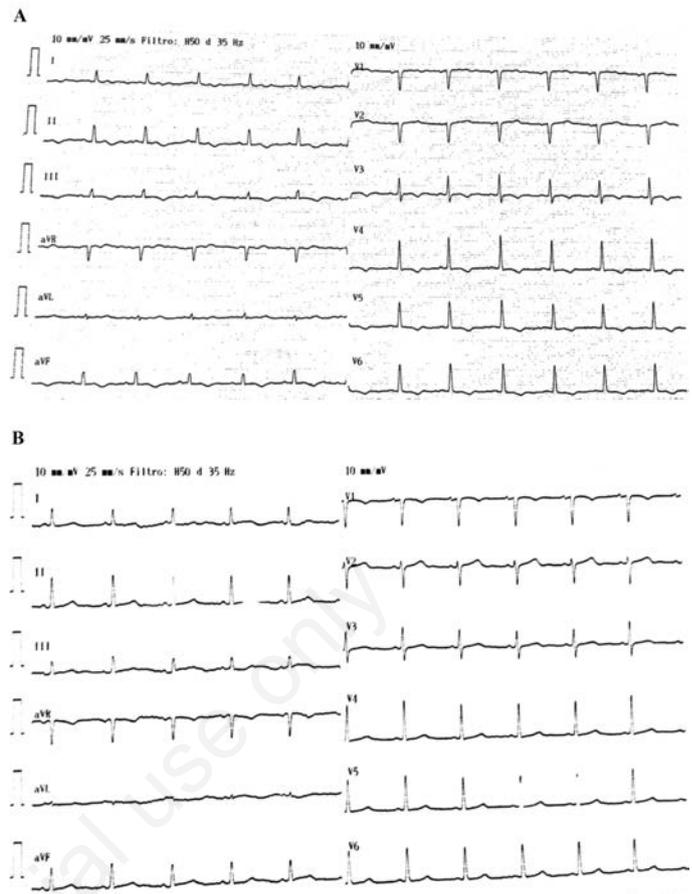
## Videos

See the following videos available online:

- Video 1: Two-dimensional parasternal long axis view showing bi-leaflet mitral valve prolapse associated with basal posterior systolic curling and mitral annular disjunction; a moderate degree of global systolic left ventricular dysfunction is also evident.
- Video 2: Two-dimensional parasternal long axis view obtained at three months after successful mitral valve repair showing disappearance of basal posterior systolic curling of the left ventricle along with absence of abnormal mitral annular disjunction and improvement of global left ventricular systolic function.



**Figure 1.** A) Basal 12-leads electrocardiogram registered some time before cardiac surgery showing normal sinus rhythm with premature ventricular beats (PVB) (isolated and coupled) with a right bundle branch block (RBBB) + left axis deviation morphology. B) Basal 12-leads electrocardiogram registered at the same time showing a short self limiting run of ventricular tachycardia with RBBB morphology and different axis deviation.



**Figure 2.** A) Basal 12-leads electrocardiogram of the patient at hospital entry before surgery showing normal sinus rhythm with negative T waves present on inferior and lateral leads. B) Basal 12-leads electrocardiogram registered 3-months after surgical repair of the mitral valve showing disappearance of T wave inversion previously evident in inferior and lateral leads.

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