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CASE PRESENTATION

# Malignant arrhythmia in mitral valve prolapse: is surgical management enough?

## *Arritmia maligna en prolapso de la válvula mitral: ¿es suficiente el tratamiento quirúrgico?*

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## Abstract

*Mitral valve prolapse is a well-known condition that is generally benign, but can be associated with cardiac arrhythmias, particularly malignant ventricular arrhythmias and sudden cardiac death. This association and its outcome have been described in medical literature, but the low incidence leads to a lack of evidence regarding its management and stratification. We present the case of a young woman whose initial manifestation was syncope, followed by severe mitral regurgitation and frequent ventricular extrasystoles. She underwent surgical repair with mitral valve plasty and cryoablation of the anterior papillary muscle. The clinical outcome was favorable, as evidenced by an improvement in symptoms and control of the arrhythmia burden.*

***Keywords:*** *Mitral valve prolapse. Arrhythmic mitral valve prolapse. Premature ventricular contractions. Ventricular arrhythmia. Cryoablation. Surgical ablation.*

## Resumen

*El prolapso de la válvula mitral es una enfermedad ampliamente conocida, la cual es benigna en la mayoría de casos; sin embargo, puede estar en asociación con alteraciones del ritmo cardiaco, específicamente arritmias ventriculares malignas y muerte cardiaca súbita. Pese a que esta asociación y su desenlace están descritos en la literatura médica, su baja incidencia conduce a la falta de evidencia en cuanto a su manejo y estratificación. Se presenta el caso de una mujer joven, cuya manifestación inicial fue síncope, con posterior desarrollo de insuficiencia mitral grave, con hallazgo de extrasístoles ventriculares frecuentes, en quien se realizó tratamiento quirúrgico con plastia mitral y crioablación del músculo papilar anterolateral. La evolución clínica fue satisfactoria, ya que se evidenció mejoría de la sintomatología y control de la carga arrítmica.*

***Palabras clave:*** *Prolapso de la válvula mitral. Prolapso arrítmico de la válvula mitral. Contracciones ventriculares prematuras. Arritmia ventricular. Crioablación. Ablación quirúrgica.*

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## Introduction

Mitral valve prolapse is one of the most common valve diseases. It may be asymptomatic for a long period of time and may present with a variety of clinical signs and symptoms ranging from mild and non-arrhythmic manifestations to the potential onset of severe mitral regurgitation with left ventricular dilation and, in specific cases, ventricular arrhythmias which may be associated with sudden cardiac death, with a 1.2 to 1.4% rate of occurrence per year[1,2]. These characteristics give this disease great clinical interest.

The pathophysiological substrate to explain fatal ventricular arrhythmias may be related to anatomical substrates, like the onset of patchy fibrosis between the mitral valve, papillary muscles and adjacent inferobasal left ventricular myocardium and triggers linked to the stretching of papillary muscles, causing early depolarization that initiates and perpetuates the triggered activity[3]. In addition, underlying mechanisms have been described, including increased adrenergic activity, catecholamine regulation disturbances, and abnormalities in the renin-angiotensin-aldosterone system[4].

In the recent description and characterization of this condition, the different clinical studies have identified risk factors directly associated with sudden cardiac death and malignant arrhythmias in patients with mitral valve prolapse, including female sex, prolapse of both valve leaflets, mitral annulus abnormalities (like dilation and disjunction), electrocardiographic repolarization abnormalities (specifically in the inferior-lateral leads), frequent premature ventricular contractions and myocardial fibrosis in the papillary muscles[1,5].

The case presented below shows the benefit of appropriate stratification in a symptomatic patient with high risk factors, in whom early surgical intervention not only improved her quality of life, but also reduced the arrhythmic burden. This case provides an avenue to inquire about the protective role of surgery in malignant arrhythmias due to mitral valve prolapse and highlight the benefit of multidisciplinary clinical follow up.

## Clinical case

We present the case of a previously healthy 38-year-old woman who initially presented due to syncope, with a finding of mitral valve prolapse of both leaflets and mild to moderate regurgitation, for whom clinical monitoring was initially considered. On follow up after her pregnancy, she had increased clinical signs of heart failure and echocardiographic progression of the mitral regurgitation from moderate to severe, with prolapse of both leaflets (predominantly the posterior leaflet) associated with 8 mm annular disjunction, the pickelhaube sign and left cavity dilation with 60% ventricular function (Fig. 2C and E). Multidisciplinary stratification was done, including 24-hour Holter monitoring which showed frequent ventricular extrasystoles with two predominant morphologies, including right ventricular outflow tract and posteromedial morphologies, with an arrhythmic burden of 11%, as well as frequent supraventricular extrasystoles (Fig. 1). In addition, cardiac magnetic resonance imaging (Fig. 2A) showed myocardial linear late gadolinium enhancement between the basal segment of the anterolateral wall and the papillary muscles. Based on her clinical exam, progression and imaging findings, she was referred for surgery.

Surgery revealed (Fig. 2B) a mitral valve with degenerative changes suggestive of Barlow's disease, prolapse of both leaflets, predominantly P2, and disjunction of the posterior mitral annulus. In addition, fibrotic foci were found on the posteromedial and anterolateral papillary muscles, as was seen initially on cardiac magnetic resonance imaging.

Intraoperative cryoablation was performed on the described papillary muscles for 120 seconds, (anteriorly, posteriorly and at their tips), guided by visual inspection of the fibrosis. The mitral valve was repaired through a triangular resection of P2 and annuloplasty with a Physio II 36 mm mitral ring. An intraoperative transesophageal echocardiogram showed a repaired mitral valve with no residual regurgitation.

The postoperative echocardiogram revealed a 40% ejection fraction and a normally functioning mitral repair with no residual regurgitation (peak velocity 0.9 m/s; mean gradient: 2 mmHg) (Fig. 2D) as well as a resolved pickelhaube sign (Fig. 2F). Further, the follow up Holter three days after the procedure reported ventricular ectopy, a reduced arrhythmic burden at 8%, couplet episodes and one idioventricular rhythm episode lasting four beats.

The patient continued to be followed by the multidisciplinary team together with the electrophysiology service, finding a sinus rhythm with ventricular extrasystoles, with an arrhythmic burden of 10% and couplets on 72-hour Holter monitoring follow up at six months. In addition, an electrocardiogram reported ventricular extrasystoles with a right bundle branch block morphology, possibly originating from the outflow tract; this focus was not seen prior to surgery, did not improve with the surgical intervention and was therefore probably not related to the mitral valve

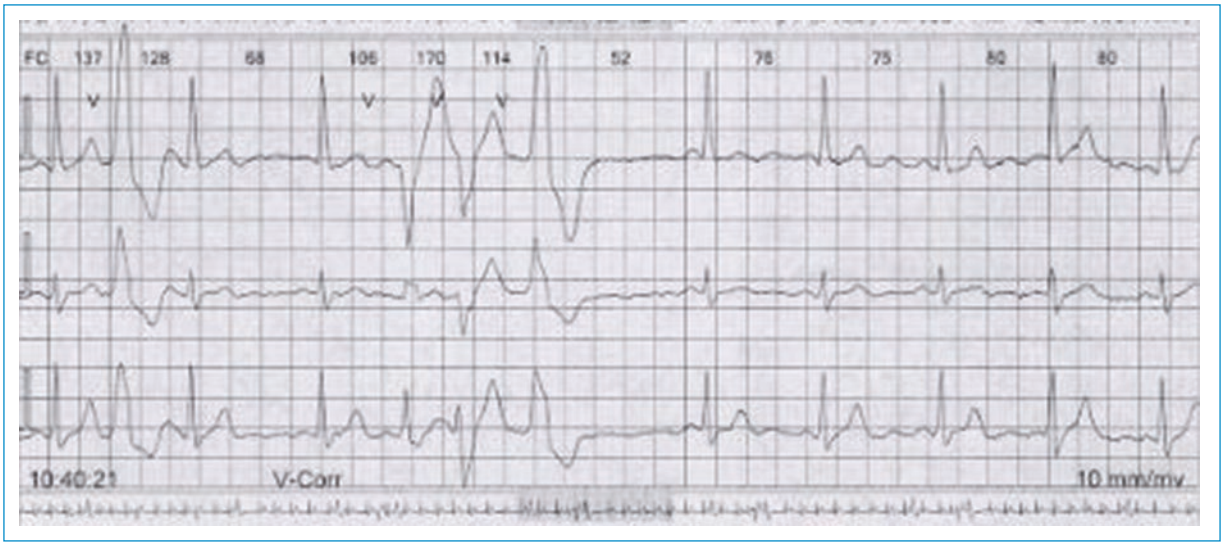


**Figure 1.** Presurgical Holter showing frequent ventricular extrasystoles with two morphologies, as well as frequent supraventricular extrasystoles.

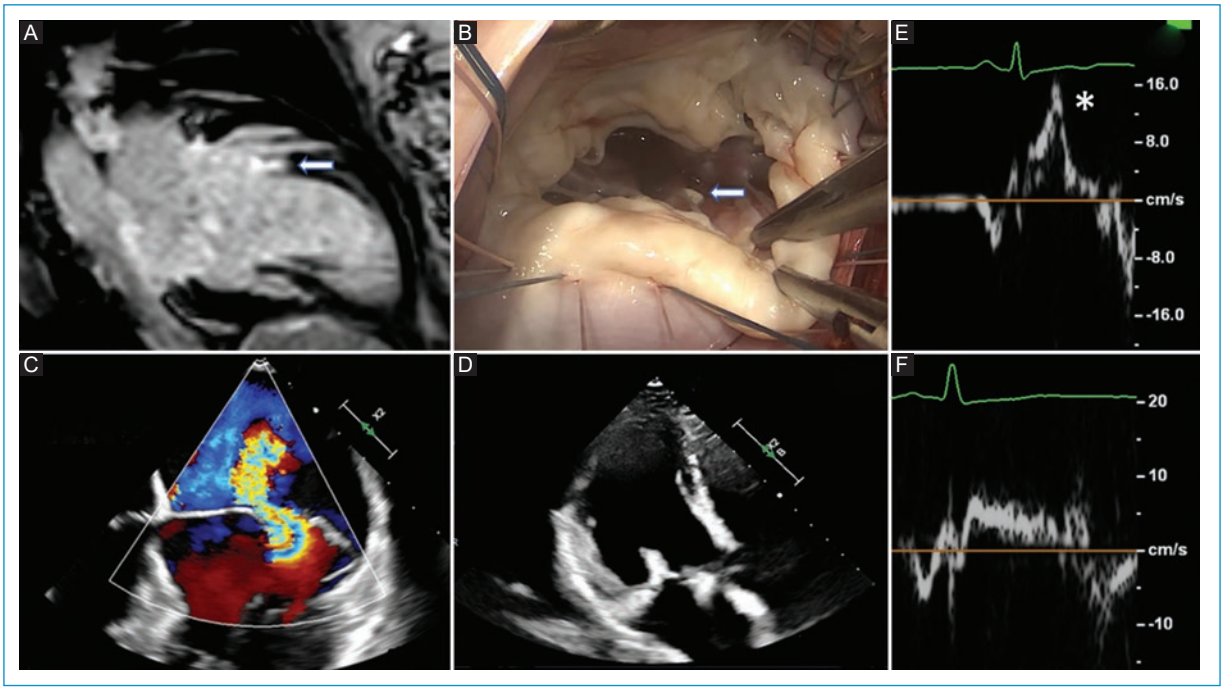


**Figure 2. A:** cardiac magnetic resonance imaging showing late gadolinium enhancement in the posteromedial papillary muscle. **B:** macroscopic correlation with the area of fibrosis in the head of the posteromedial papillary muscle. **C:** mitral valve prolapse associated with mitral annulus *curling* and disjunction. **D:** mitral valve repair with an adequate coaptation surface. **E:** preoperative pickelhaube sign. **F:** resolution of the pickelhaube sign after mitral valve repair.

prolapse. Due to these findings, she required percutaneous modulation of the arrhythmogenic substrate by electrophysiology, resulting in complete resolution of the arrhythmic burden.

## Discussion

This arrhythmic phenotype of mitral valve prolapse is closely related to mitral annular disjunction, due to its pathophysiological relationship with local and

progressive fibrosis of the papillary muscles and the inferolateral wall of the left ventricle[6]. While mitral valve prolapse and mitral annular disjunction may coexist or be independent entities, it is clear that mitral annular disjunction is a predictor of the risk of arrhythmias[7]. The prevalence of mitral valve prolapse and mitral annular disjunction ranges from 20 to 58%. Although not all types of mitral annular disjunction are associated with ventricular arrhythmias, those that are are directly concordant with advanced myxomatous disease.

Miller et al.[6] described the mechanisms that interact with the development of sustained ventricular arrhythmias and sudden death in patients with mitral valve prolapse: a) a substrate (scar), b) a trigger (automaticity, reentry and triggered activity), and c) a transient modulator (a hyperadrenergic state, hemodynamics, and electrolytes). Other strongly associated factors include ST segment changes, which can be understood as changes secondary to a structural abnormality, and marked leaflet redundancy[6]. The pickelhaube sign has been described as a new echocardiographic sign and a sign of early electrical dysfunction during electrophysiological tests for arrhythmic mitral valve prolapse syndrome. It is defined as a high-velocity mid-systolic acceleration spike caused by mechanical traction on the posteromedial papillary muscle by the prolapsing myxomatous leaflets leading to abrupt movement of the adjacent posterobasal left ventricular wall towards the apex. It represents the mechanical stress on the valvular and subvalvular apparatus caused by mitral prolapse and mitral annular disjunction[8,9].

The survival of patients with mitral valve prolapse and ventricular arrhythmias is directly related to the complexity of the premature contractions and the arrhythmic burden, as these are related to the probability of sudden death[10]. However, only 10% of patients with mitral valve prolapse have severe arrhythmias, predominantly those who have left atrial and ventricular dilation and degenerative myxomatous disease[11].

The available evidence to date has reported a relationship between the severity of the ventricular arrhythmias in these cases and the related fatal outcomes. However, the role of arrhythmogenic focus ablation and its timing (before, during or after surgery) in preventing the risk of malignant arrhythmias is unclear, as well as whether mitral valve repair is sufficient[10]. El-Ashmawi et al.[12] published a case series in which patients with malignant arrhythmic mitral valve prolapse underwent mitral valve repair with cryoablation towards the fibrosis foci, and showed a short-term reduction of the arrhythmic burden. However, we cannot affirm that our patient had a significant reduction in the arrhythmic burden after cryoablation but can state that her symptoms improved with mitral repair.

The subgroup of patients with mitral valve prolapse, annular disjunction and ventricular arrhythmias requires stratification and multidisciplinary assessment by an extended Heart Team (cardiology, cardiovascular surgery, electrophysiology and radiology) to identify those who would benefit from early surgical treatment, direct intraoperative ablation and multidisciplinary follow up, as these patients, like the case we have presented, may require percutaneous interventions. Currently, Holter monitoring, cardiac magnetic resonance imaging and echocardiography are used for stratification to identify risk factors, determine the arrhythmic burden and correlate scars with potential arrhythmogenic substrates, and establish the treatment plan.

## Conclusion

In this report of a patient with mitral valve prolapse, we found improved symptoms when cryoablation of the arrhythmogenic substrate in the late gadolinium enhancement sites was combined with mitral valve repair. However, so far, surgical cryoablation does not ensure a reduced risk of sudden death, despite prior indications to this effect. Therefore, this subgroup of patients requires comprehensive multidisciplinary stratification, assessment and management before and after surgery. In addition, a randomized study is needed to determine if intraoperative muscle ablation reduces the arrhythmic burden, compared with isolated mitral repair.

## Ethical considerations

Approved by the Research Ethics Committee at La Cardio.

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## Conflicts of interest

The authors declare no conflicts of interest.

## Ethical disclosures

**Human and animal protection.** The authors declare that no experiments were conducted on humans or animals in the course of this study.

**Data confidentiality.** The authors declare that they have followed their workplace protocols for publishing patient data.

**Right to privacy and informed consent.** The authors have obtained informed consent from the patients and/or subjects referred to in the article. The corresponding author is in possession of this document.

**Use of artificial intelligence to generate text.** The authors declare that they have not used any type of generative artificial intelligence for writing this manuscript or creating figures, graphs, tables or their respective captions or legends.

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