Recurrent premature ventricular contraction–induced ventricular fibrillation and resuscitated sudden death in a 26-year-old pregnant woman with bileafllet mitral valve prolapse

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Introduction
Mitral valve prolapse has been identified as a risk factor for sudden cardiac death (SCD) with an annual rate of 0.2%–0.4%, which is approximately 2- to 4-fold higher than that of the general population.1,2 Recently, a “malignant” variant has been identified in patients with bileafllet prolapse experiencing a higher rate of ventricular arrhythmias.3–6 Supraventricular arrhythmias are the most common and remaining ventricular arrhythmias rather uncommon in pregnancy.7

SCD due to sustained ventricular tachycardia (VT) and ventricular fibrillation (VF) is exceedingly rare in pregnancy. It is most commonly precipitated by other systemic illnesses or complications due to pregnancy.8 We present a case of a young 13 weeks pregnant woman with bileafllet prolapse who presented with premature ventricular contraction (PVC)–induced VF with resuscitated SCD. She was controlled with medication and defibrillator implant and delivered successfully at term. The patient had a second pregnancy also with increased ventricular arrhythmia burden controlled medically. After the delivery of her second child, the patient experienced increased burden of single monomorphic PVCs that recurrently induced nonsustained VF that were successfully mapped and ablated from the Purkinje network and posteromedial papillary muscle.

Case report
A 26-year-old woman was transferred to our hospital after an episode of resuscitated SCD. She was a physical education teacher who was 13 weeks pregnant at the time of her cardiac arrest. She was noted to have an abrupt syncope while teaching a gym class. Cardiopulmonary resuscitation was initiated, and an automated external defibrillator (AED) was used. The initial rhythm was noted to be VF, and an appropriate shock was applied. She regained consciousness and reverted to normal sinus rhythm. Seconds later, the patient had a recurrent syncope and recurrence of PVC-induced VF. Cardiopulmonary resuscitation was reinitiated, and the patient had 4 more rounds of successful defibrillation of PVC-induced VF by the AED. She developed atrial fibrillation and continued to have some PVCs (Figure 1). She converted to normal sinus rhythm and was transferred to our hospital for further care. The patient did not have any further PVCs on telemetry in 48 hours. The echocardiogram showed bileafllet mitral valve prolapse with moderate mitral regurgitation and a normal ejection fraction. Cardiac catheterization and cardiac magnetic resonance imaging (MRI) were deferred because of risks of exposure during pregnancy. A single-chamber transvenous defibrillator (Boston Scientific, Marlborough, MA) was placed via the left cephalic approach without any complications. Both the patient and the fetus remained hemodynamically stable and were discharged home. Holter monitoring performed 1 month after discharge showed <0.2% burden of PVCs and no further episodes of PVC-induced VF.

During the second trimester, the patient began to have palpitations secondary to monomorphic PVC-induced nonsustained VT. Sotalol was initiated and titrated up to 120 mg twice daily as an inpatient. The patient delivered a healthy baby boy at full term without any complications. She had a reduction in PVCs without significant episodes of VT after delivery. Genetic testing with “sudden death panel” (Transgenomic) was negative. Cardiac MRI showed bileafllet mitral valve prolapse with moderate to severe mitral regurgitation and a normal ejection fraction. Late gadolinium enhancement (LGE) of the papillary muscles, right ventricular (RV) outflow tract, or epicardium was not seen in her cardiac MRI. The MRI protocol used was the “viability protocol,”

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which is the standard protocol used at the University of Pennsylvania. The range of thickness varies by the sequence acquisition from 4 to 8 mm; for the cine MRI imaging sequence, the slice thickness was 8 mm. Eventually she began to have symptoms of marked sinus bradycardia and the sotalol dose was decreased to 40 mg daily. At the age of 29, she was pregnant again and experienced an increase in the total burden of monomorphic PVCs inducing short nonsustained episodes of VT. Her sotalol dose was titrated up to 120 mg twice daily, with improvement in symptoms and suppression of PVCs and VT. The patient delivered her second healthy baby boy without any complications.

One year after delivery, she presented again with recurrent episodes of symptomatic PVC-induced nonsustained VT. Her Holter monitor at that time showed 11% PVCs, some triggering nonsustained VT. Her human chorionic gonadotropin urine test at that time was negative. Our patient elected for catheter ablation. Mapping and ablation were performed using a 3-dimensional electroanatomic mapping system with an intracardiac echocardiography (ICE) catheter (CartoSound software and SoundStar catheter, Biosense Webster Inc., Diamond Bar, CA), a 3.5-mm open irrigated-tip mapping, and an ablation catheter (NaviStar ThermoCool SmartTouch, Biosense Webster). A 3D electroanatomic map was created with

**KEY TEACHING POINTS**

- Bileaflet mitral valve prolapse is an uncommon but increasingly frequently recognized risk factor for sudden death due to premature ventricular contraction (PVC)-induced ventricular fibrillation (VF).
- Pregnancy may cause acute exacerbation of ventricular arrhythmias or precipitate sudden death in this population.
- Intracardiac echocardiography can be a sensitive tool for diagnosing abnormal papillary muscle structures and guiding ablation that may be the site of origin of PVC-induced VF; it remains to be seen if prospective identification of abnormal papillary muscle by intracardiac echocardiography could identify patients with bileaflet prolapse at risk for sudden death before their index event.
- Rapid atrial pacing or atrial arrhythmias/atrial fibrillation, as in this case, can be temporarily “protective” from recurrent PVC-induced VF or ventricular tachycardia/VF storm.

![Image](image)

**Figure 1** A and B: Recurrent premature ventricular contraction–induced ventricular fibrillation. C: “Protective” effect of spontaneous atrial fibrillation. Note that premature ventricular contractions are no longer able to induce ventricular fibrillation.
and with 10 without recurrence at 1-hour waiting period without sedation with initial suppression and then recurrence. After multiple at-sites were ablated along the base of the papillary muscle episodes of nonsustained rapid polymorphic VT. Multiple tion at this site with a contact force of 15 g and 25 W induced were mapped to the LV posteromedial papillary muscle. Ablation of this site terminated PVCs locally. In addition, early sites and found to be relatively normal (Figure 3B). Ablation of created a voltage map of the RV and LV in sinus rhythm in 24 hours) but without symptoms of PVCs or VT. The patient had rare PVCs of different morphologies (Figure 3A). The papillary muscles were noted to have an abnormal white and enlarged “moth eaten” appearance consistent with fibrosis and scarring (Figure 2). We recorded a voltage map of the RV and LV in sinus rhythm and found to be relatively normal (Figure 3B). Ablation of this site terminated PVCs locally. In addition, early sites were mapped to the LV posteromedial papillary muscle. Ablation at this site with a contact force of 15 g and 25 W induced episodes of nonsustained rapid polymorphic VT. Multiple sites were ablated along the base of the papillary muscle with initial suppression and then recurrence. After multiple attempts with contact force of 15 g and 30 W, PVCs terminated without recurrence at 1-hour waiting period without sedation and with 10 μg of isoproterenol. At 2 months of follow-up, the patient had rare PVCs of different morphologies (<100 in 24 hours) but without symptoms of PVCs or VT.

Discussion
While palpitations and single PVCs are common in pregnancy, cardiac arrest due to primary arrhythmia is distinctly uncommon. It was fortuitous that our patient was around trained professionals and an accessible AED. The AED recordings revealed recurrent PVC-induced VF/VF storm. To our knowledge, this is the first case of PVC-induced VF in a pregnant woman with a “malignant variant” of bileaflet prolapse presenting with SCD. The apparent increase in frequency of PVCs and subsequent VT in this patient during both pregnancies suggests that hormonal factors may make pregnancy particularly risky in patients with this condition.

The development of spontaneous atrial fibrillation after repeated appropriate AED shocks was also coincidentally lifesaving in this patient (Figure 1). The rapid rate and irregularity of rhythm in this case prevented progression of her arrhythmia to recurrent VF. Although possibly coincidental, it appears that the rapidity and irregularity prevented a predictable coupling interval or interrupted a reentrant process within the Purkinje network to induce VF. This reflects some of the common practices among electrophysiologists to increase heart rate with medications such as Isuprel in an attempt to suppress recurrent VF. It would be interesting to see if an automatic device algorithm detecting recurrent PVC-induced VF would be able to suppress VF storm by rapidly pacing the atrium with various R-R intervals mimicking atrial fibrillation, as seen in this case.

Although generally considered safe in pregnancy, cardiac MRI was not recommended during the initial hospitalization because of fetal risks before 22 weeks of gestation. After her delivery, cardiac MRI did not show any evidence of LGE. A standard viability protocol was used in which slice thickness varies from 4 to 8 mm with a cine MRI imaging sequence thickness resolution of 8 mm. LGE of the papillary muscles is not uncommon and associated with ventricular arrhythmias. This case is consistent with a previous case series of 14 patients, which did not demonstrate LGE in the papillary muscles at the PVC site of origin. We created a voltage map of the LV in sinus rhythm and demonstrated relatively normal voltage throughout the LV and RV including the papillary muscles (Figure 3B). The papillary muscle was abnormal on ICE. There is a lack of a standard criterion to describe such findings. The papillary muscles were enlarged and had a “moth eaten” appearance with areas of brightness, generally unseen on ICE during ablation procedures. We would be curious to know if patients with this malignant variant of bileaflet prolapse could benefit from ICE screening to prevent the incidence of significant ventricular arrhythmias or arrests. It is possible that the area of fibrosis is so small that it is below the resolution of cardiac MRI; in addition, a voltage map may not be able to identify microscopic areas that are below the endocardial surface; however, ICE may be a viable option to detect and target potential triggers for PVC-induced VF.

Although increasingly recognized as a trigger for sudden death, the overall prevalence of PVC-induced VF from the papillary muscles in the setting of bileaflet prolapse remains relatively uncommon. We would propose to create a multi-institutional intracardiac image bank from patients undergoing ablation to see if a common criterion could be established to identify those with a true “high risk” within the population of patients with mitral valve prolapse.

Conclusion
Mitral valve prolapse is increasingly recognized as a cause of SCD due to PVC-induced VF. Ventricular arrhythmias are exacerbated in pregnancy. ICE can identify abnormal papillary muscle structures otherwise not identified with voltage mapping or with cardiac MRI. Rapid atrial fibrillation is potentially protective in recurrent PVC-induced VF.
Figure 3  
A: Twelve leads of premature ventricular contraction morphology and site of early activation and successful termination of premature ventricular contraction with ablation. 
B: Electroanatomic map created during sinus rhythm demonstrating relatively normal myocardial voltage with earliest sites of PVC origin overlay.
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