Received: 1 May 2019 | Revised: 31 May 2019 | Accepted: 17 June 2019

DOI: 10.1002/joa3.12220

CASE REPORT

Journal of Arrhythmia Open Access WILEY

Nearly fatal ventricular arrhythmia following pacemaker implantation in a young female with complete heart block

Hussam Ali MD, FESC, FEHRA, FAIAC[1] | Pierpaolo Lupo MD[1] | Sara Foresti MD[1] | Guido De Ambroggi MD[1] | Riccardo Mantovani MD[1] | Carmine De Lucia MD[1] | Ilaria Passarelli MD[1] | Luigi De Ambroggi MD, FESC[2] | Riccardo Cappato MD, FESC, FHRS[1]

[1]Arrhythmia & EP Research Center, IRCCS Humanitas Research & University Hospital, Rozzano (Milan), Italy

[2]University of Milan, Milan, Italy

Correspondence
Hussam Ali, MD, FESC, FEHRA, FAIAC, Arrhythmia and EP Research Center, IRCCS Humanitas Research & University Hospital, Via A. Manzoni 56 – 20089 - Rozzano (Milan), Italy.
Email: hussamali.ep@gmail.com

Abstract

This case concerns a 24-year-old female who developed malignant ventricular tachyarrhythmia a few weeks after pacemaker implantation for complete heart block. Apparently, right ventricular pacing caused significant repolarization abnormalities in both native and paced rhythms with marked QT prolongation and substantial electrical instability. This case highlights other intriguing phenomena in the puzzle of cardiac repolarization and how pacing therapy may alter this complex process providing arrhythmic substrate in vulnerable subjects. Though such arrhythmic events are clinically rare, vulnerable patients or with suspected myocardial disease that may cause QT prolongation should be carefully followed in the course of pacing therapy.

KEYWORDS

complete heart block, long QT, pacemaker, ventricular repolarization, ventricular tachyarrhythmia

1 | INTRODUCTION

Ventricular repolarization is a complex process that may be altered by nonphysiological ventricular depolarization and T wave changes are well known after prolonged ventricular pacing "electrical memory".[1] Though cardiac pacing could be life-saving in many conditions, it may induce malignant proarrhythmic effects in vulnerable subjects as in our patient.

2 | CASE REPORT

A 24-year-old female patient with known complete heart block underwent uneventful dual-chamber pacemaker implantation due to initial functional limitation and prolonged pauses. She had no family history of cardiomyopathy, conduction defects, neuromuscular disorders, long QT syndrome, or sudden cardiac death. Echocardiogram and previous cardiac resonance excluded significant structural heart disease. Basal ECG (Figure 1A) showed sinus rhythm, complete heart block and escape junctional rhythm at 47 bpm, with a normal QTc (407 ms according to the Bazett formula). The device was programmed in DDD modality at a lower-rate of 50 bpm, and an ECG before discharge (Figure 1B) showed sinus rhythm at 53 bpm, paced-QRS in atrial-tracking modality with a normal QTc (404 and 423 ms, calculated according to the Bogossian and Wang Formulas, respectively).[2,3]

A few weeks following implantation, the patient was admitted to the emergency department for syncope that occurred at rest and was associated with convulsions. Physical examination was unremarkable and vital signs were normal. At initial evaluation, an epileptic crisis was suspected as the underlying mechanism. However, a 12-lead ECG (Figure 1C) revealed sinus rhythm and paced-QRS with

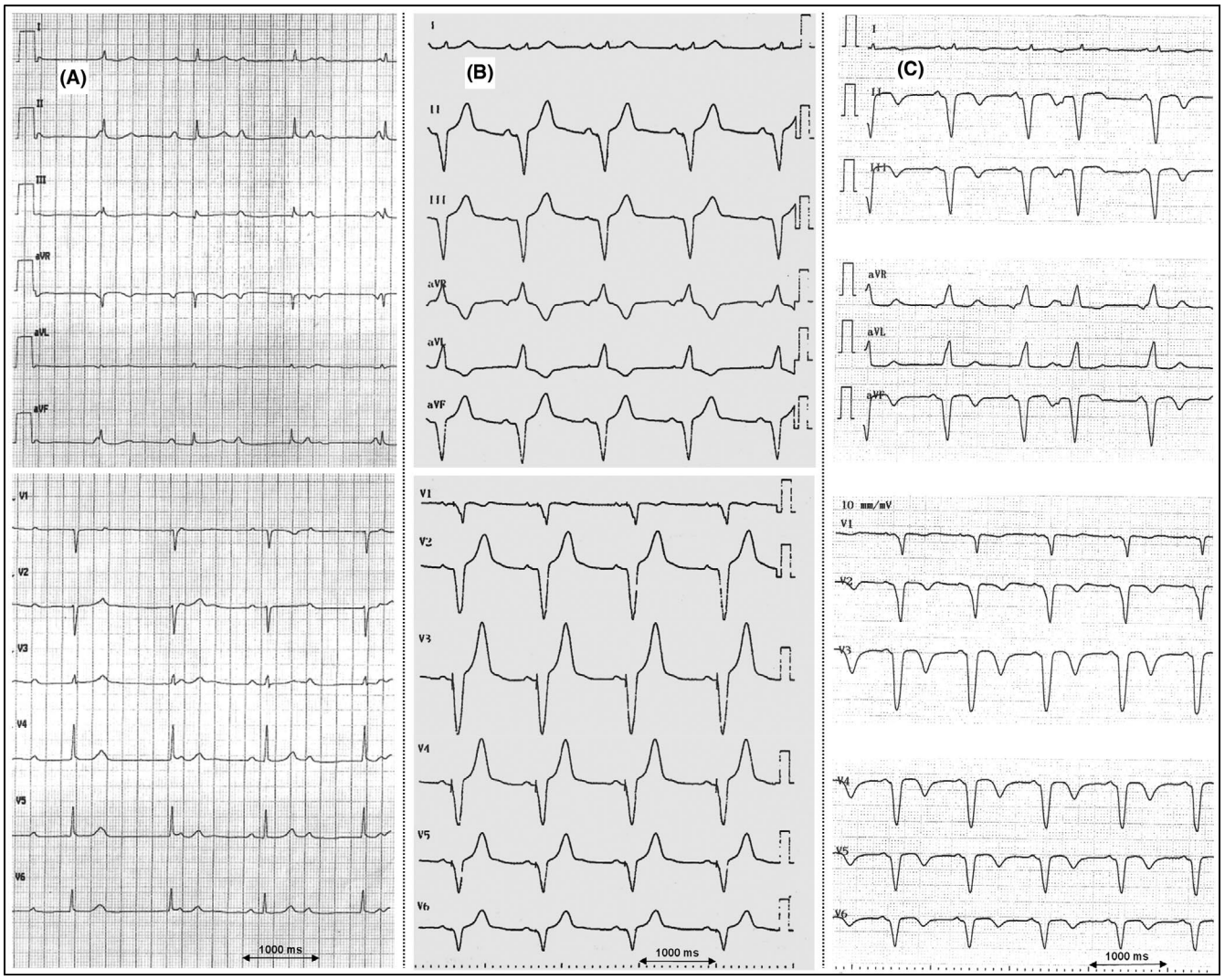

FIGURE 1 A, Basal 12-lead ECG showing sinus rhythm, complete heart block, and escape junctional rhythm with normal repolarization. B, 12-lead ECG the day after pacemaker implantation showing sinus rhythm and ventricular pacing in DDD modality. C, 12-lead ECG on admission after the syncopal episode revealing sinus rhythm, paced-QRS with diffuse T wave inversion and QT prolongation

a slight increment in duration (136 ms vs. 130 ms acutely after implantation) and showing T wave inversion in the inferior and chest leads with a relatively prolonged QTc (472 and 493 ms calculated according to the Bogossian and Wang Formulas, respectively). Moreover, pacemaker interrogation revealed a corresponding prolonged episode of ventricular fibrillation or polymorphic ventricular tachycardia, triggered by a late couple of premature ventricular beats, and lasting up to 2 minutes with spontaneous termination (Figure 2A). Remarkably, pacing inhibition unmasked native escape junctional rhythm at 62 bpm and diffuse giant negative T waves with a markedly prolonged QTc (602 ms) and T wave alternans indicative of substantial electrical instability (Figure 2B). Blood tests, including blood cell count, CRP, ESR, electrolytes, myocardial enzymes, D-Dimer, and thyroid function, were all within normal limits. There was no pharmacological therapy that may cause QT prolongation. Echocardiogram showed normal biventricular function and no pericardial effusion. A cardiothoracic angio-CT did not show any relevant coronary lesions or signs of pulmonary embolism. The patient had no neurological impairment and was admitted to the intensive care unit for strict monitoring and put on beta-blocker therapy. Subsequent ECG telemetry did not document any ventricular arrhythmias. After detailed discussion with the patient and her family, the clinical decision was to upgrade the pacing system to a dual-chamber defibrillator, and the apical right ventricular lead was replaced with a defibrillation lead positioned at the interventricular septum.

3 | DISCUSSION

The exact mechanism of repolarization abnormalities and the malignant ventricular arrhythmia in our patient is unclear and secondary causes of long QT could not be identified. Repolarization abnormalities and QT prolongation may be caused by medications, electrolyte abnormalities, myocardial ischemia, hypothyroidism,

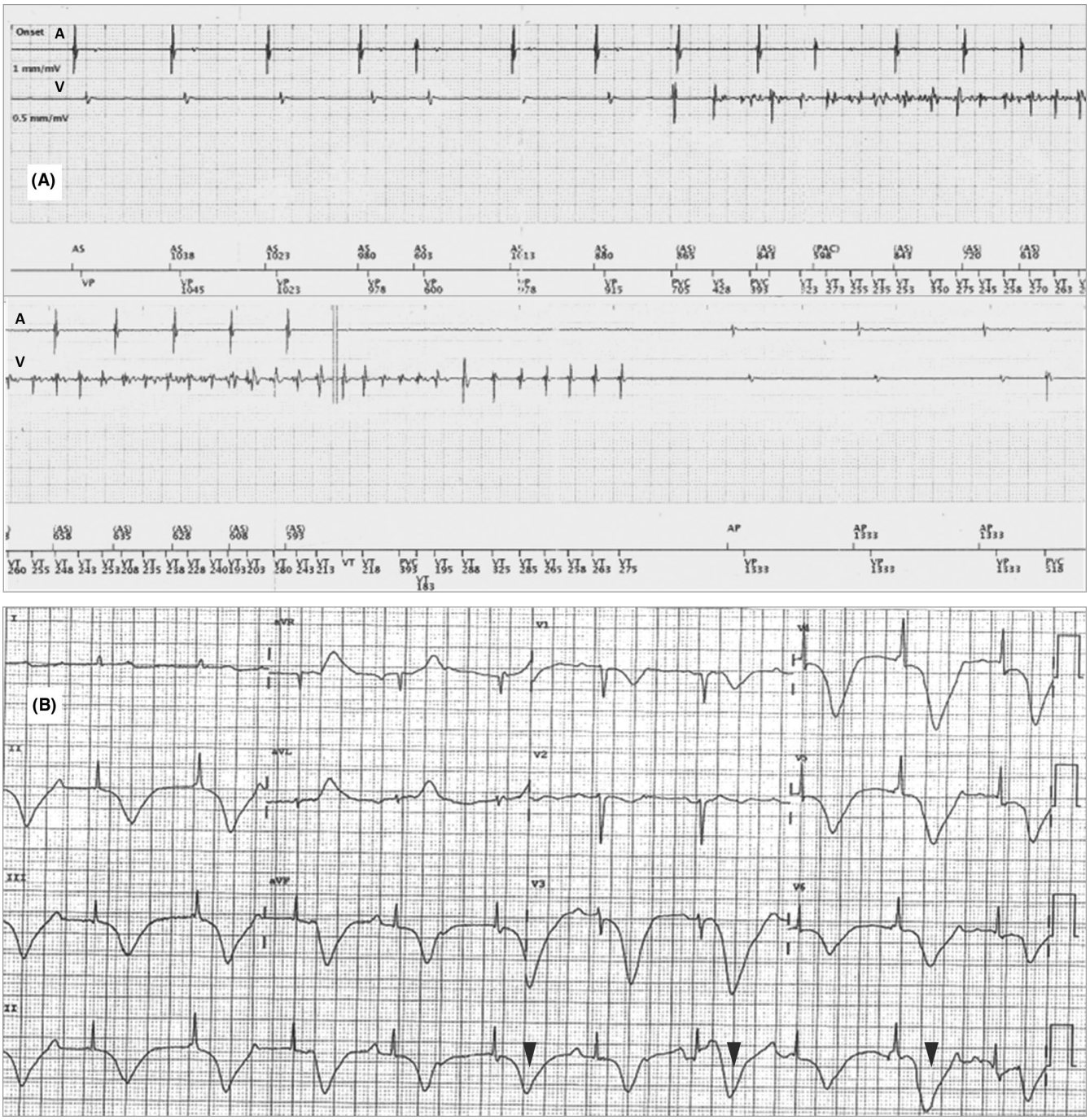

FIGURE 2 A, Noncontinuous pacemaker recording of bipolar atrial and ventricular electrograms showing the onset (upper part) and offset (lower part) of the arrhythmic event. The double vertical lines in the lower part indicate resumption of recording before arrhythmia termination. B, 12 lead-ECG of native rhythm after the syncopal episode, obtained during pacemaker deactivation, showing sinus rhythm, complete heart block and escape junctional rhythm with profound repolarization abnormalities including diffuse giant negative T waves, prolonged QTc and a T-wave alternans pattern (the arrows)

pulmonary embolism, intracranial disease, and structural heart disease. Though coronary vasospasm cannot be excluded with a normal coronary CT, the patient denied any angor or equivalent symptoms, particularly before the arrhythmic event. The new-onset of complete heart block in such young patient should raise the suspicion of underlying structural heart disease, but abnormal findings were absent at cardiac imaging modalities including echocardiogram, cardiac CT and previously performed cardiac resonance. However, some rare cardiomyopathies as cardiac sarcoidosis may require further diagnostic studies as positron-emission tomography. Of note, the arrhythmic episode was not triggered by a prolonged pause or pacemaker malfunction that was also excluded at device interrogation. Furthermore, fluoroscopic check showed stable and normal position/motion of the ventricular lead. Accordingly, the arrhythmic

episode may be favored by the applied pacing therapy due to the temporal interrelation between events. Moreover, before pacemaker implantation, the patient never had major symptoms (syncope or presyncope), and a long QT interval or complex ventricular arrhythmias were not observed through the long-term and strict follow-up over the last 5 years.

Ventricular pacing is known to affect repolarization, mainly due to prolonged depolarization (QRS widening), and T wave changes may be observed after prolonged ventricular pacing (cardiac electrical memory).[1] Hence, the observed repolarization alterations (Figure 2B) may be, at least to in part, a reflection of cardiac memory that was provoked by the abrupt repolarization change after inhibition of ventricular pacing and restoration of the native rhythm. In this patient, there was no 12-lead ECG available just before the arrhythmic event, and thus it is uncertain which occurred first (i.e., electrocardiographic changes or the ventricular arrhythmia). However, repolarization abnormalities did persist at ECG recordings through the subsequent months after this single and relatively short episode of ventricular arrhythmia, making the hypothesis of arrhythmia-induced repolarization changes less likely. Noteworthy, Takotsubo cardiomyopathy may produce similar giant negative T waves that usually recover within several months, and it is an important differential diagnosis since pacemaker implantation in a young woman can be a remarkably stressful condition. Nevertheless, normal echocardiogram and cardiac CT findings with the absence of the classical "apical ballooning" make Takotsubo unlikely as an underlying mechanism. Therefore, right ventricular pacing may have led to profound de novo repolarization abnormalities in our patient.

Assessment of the QT interval during paced rhythms can be challenging, but a few methods have been developed in clinical practice.[2,3] By using two different formulas, The QTc of the paced-QRS was substantially increased (by ~ 70 ms) several weeks after implantation when the arrhythmic event occurred. Thought the arrhythmia onset was not the typical short-long pattern of *torsades de points* in long QT, continuous ventricular pacing likely led to remarkable repolarization abnormalities manifested as prolonged QT and T wave alternans providing a vulnerable substrate for ventricular tachyarrhythmias. Importantly, the arrhythmic episode occurred at rest with a heart rate of 58 bpm excluding rate-dependent long QT, unmasked after pacemaker implantation, as a possible mechanism. Despite pacing therapy has been used to reduce the arrhythmic burden in some patients with acquired or congenital long QT,[4] the ventricular pacing itself and nonphysiologic ventricular depolarization may affect cardiac repolarization and electrical memory producing significant proarrhythmias.[5] During the upgrading procedure, the ventricular defibrillation lead was positioned at the septum, aiming to provide more physiologic activation as compared to apical pacing. Nevertheless, the long QT persisted during the follow-up despite septal ventricular pacing, excluding a pacing site-dependent effect. However, considering the temporal correlation and after exclusion of other common causes, the ventricular pacing seems to be, but still uncertain, a possible mechanism or trigger.

The device lower-rate was initially programmed at 75 bpm (DDDR modality) with the hysteresis function turned off to attenuate the long QT arrhythmic effects. The patient was proposed to undergo genetic testing at a specialized center and discharged on beta-blocker therapy. No subsequent arrhythmic events occurred at a 3-months follow-up period.

This case highlights other intriguing phenomena in the puzzle of cardiac repolarization and how pacing therapy may alter this complex process providing arrhythmic substrate in vulnerable subjects. Though such arrhythmic events are rare in clinical practice, vulnerable patients, or with suspected myocardial disease that may cause QT prolongation, should be carefully followed in the course of pacing therapy.

ACKNOWLEDGEMENT

None.

CONFLICT OF INTEREST

The authors declare no conflict of interests for this article.

ORCID

Hussam Ali https://orcid.org/0000-0002-3469-1028

REFERENCES

1. Rosenbaum MB, Blanco HH, Elizari MV, Lázzari JO, Davidenko JM. Electrotonic modulation of the T wave and cardiac memory. Am J Cardiol. 1982;50(2):213–22.
2. Bogossian H, Frommeyer G, Ninios I, Hasan F, Nguyen QS, Karosiene Z, et al. New formula for evaluation of the QT interval in patients with left bundle branch block. Heart Rhythm. 2014;11(12):2273–7.
3. Wang B, Zhang Li, Cong P, Chu H, Liu Y, Liu J, et al. A new formula for estimating the true QT interval in left bundle branch block. J Cardiovasc Electrophysiol. 2017;28(6):684–9.
4. Dorostkar PC, Eldar M, Belhassen B, Scheinman MM. Long-term follow-up of patients with long-QT syndrome treated with beta-blockers and continuous pacing. Circulation. 1999;100(24):2431–6.
5. Di Cori A, Zucchelli G, Romano S, Bongiorni MG. Proarrhythmic effect of pacing mode reprogramation in a patient with a congenital long-QT syndrome. J Cardiovasc Electrophysiol. 2013;24(11):1304–5.

How to cite this article: Ali H, Lupo P, Foresti S, et al. Nearly fatal ventricular arrhythmia following pacemaker implantation in a young female with complete heart block. *J Arrhythmia*. 2019;35:766–769. https://doi.org/10.1002/joa3.12220