An optimized long QT syndrome differentiating protocol: A new indication for wearable cardioverter-defibrillator vests

Marcin Pilawa, Jarosław Kaźmierczak, Radosław Marek Kiedrowicz

Department of Cardiology, Pomeranian Medical University, Szczecin, Poland

Correspondence to:
Radosław Marek Kiedrowicz, MD, PhD,
Department of Cardiology, Pomeranian Medical University, Powstańców Wlkp. 72, 70–111 Szczecin, Poland,
phone: +48 509 395 560,
e-mail: radkied@wp.pl

DOI: 10.33963/KP.a2022.0142

Received:
March 17, 2022

Accepted:
May 10, 2022

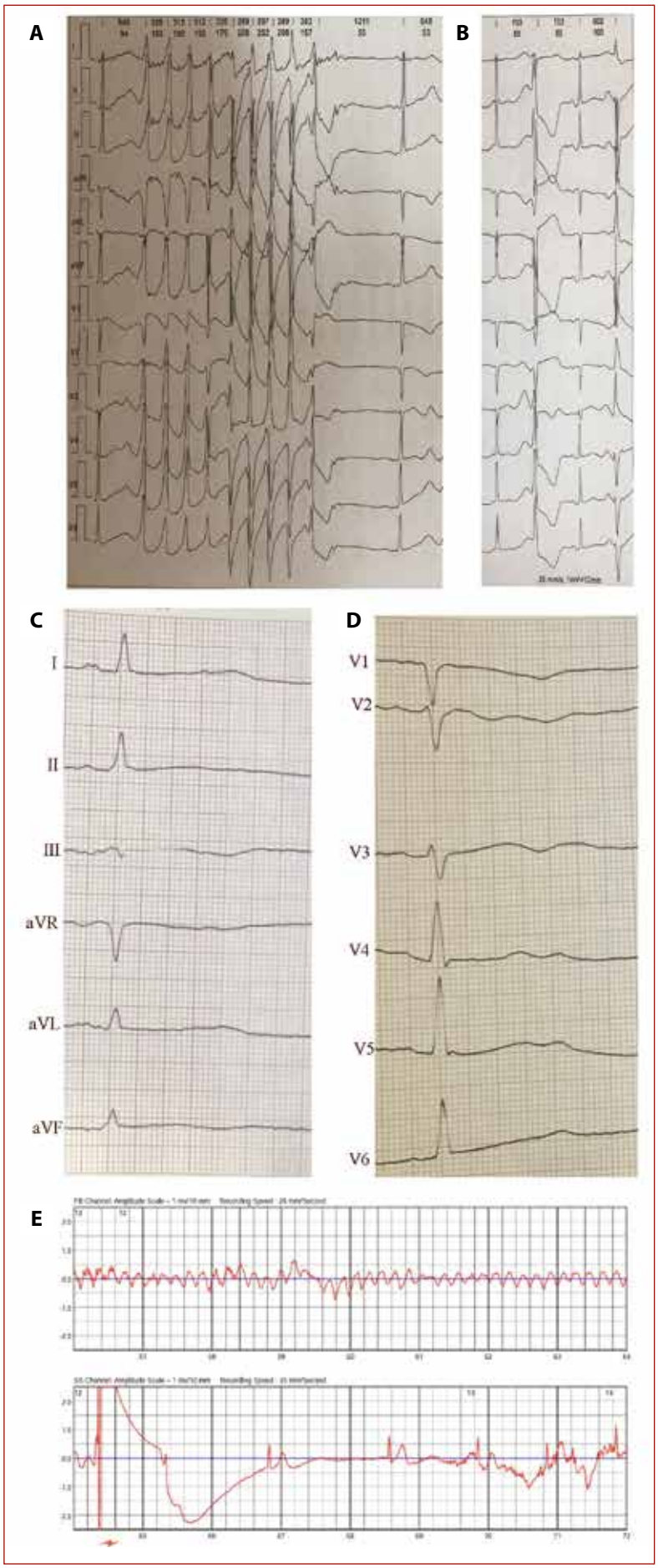
Early publication date:
June 7, 2022

A 29-year-old female diagnosed with ventricular tachycardia (VT) was transferred to our department for emergency ablation. She had been suffering from infrequent syncope since she was 21 years of age. The unexplained syncope almost always occurred after the patient suddenly awoke. Her past medical tests had excluded central nervous system, carotid artery or structural heart disease, heart rhythm disturbances, and a family history of sudden deaths. She was not taking any drugs.

Nine months after childbirth, she was admitted to another center due to a series of syncopal episodes. A Holter electrocardiographic (ECG) monitoring showed 20 300 ventricular extrasystoles (VEs) in 24 hours along with 291 non-sustained VT episodes lasting up to 17 seconds, triggered by VEs with a very short coupling interval (Figure 1A, B). She received a 9 g loading dose of amiodarone and metoprolol, resulting in complete arrhythmia suppression.

At our department, neither VEs nor VTs were found during her hospital stay and an ablation procedure was canceled. Twelve-lead ECGs showed a sinus rhythm of 60 beats per minute and a corrected QT interval (QTc) of 540 ms with low amplitude, bifid T waves in the absence of electrolyte imbalance (Figure 1C, D). Her clinical history was matched to subtype 2 long QT syndrome (LQTS). According to the European Society of Cardiology guidelines, an implantable cardioverter-defibrillator (ICD) was considered as she continued to be at risk of cardiac arrest due to QTc >500 ms [1], though in addition, drug-induced LQTS was very likely. Therefore, it was decided to discontinue amiodarone, prescribe a non-cardioselective β-blocker, and verify QTc after a 3-month washout period. During this waiting time, she was protected by using a wearable cardioverter-defibrillator vest (WCD; LifeVest 4000, Zoll, Pittsburgh, PA, US). Twelve weeks following discharge the patient was awakened suddenly by her crying child and lost consciousness. The WCD registered ventricular fibrillation (VF) which was successfully cardioverted (Figure 1E). Her QTc was not shortened, and she was finally diagnosed with LQT2 and a transvenous ICD was implanted. No VT/VF was registered on 3-month follow-up.

Inherited LQTS, which results in cardiac ion channel abnormalities, is present in 1 in every 2000 births [2]. Acquired LQTS is usually caused by drugs or electrolyte abnormalities and occurs in 2.0%–8.8% of patients who are prescribed anti-arrhythmic drugs. In particular, Vaughan-Williams classification category III antiarrhythmic drugs, including amiodarone, carry the risk of causing proarrhythmic effects due to QT prolongation [3]. In the presented case differential diagnosis included congenital and amiodarone-induced LQTS. It was presumed that QT shortening after the amiodarone washout period, along with VEs ablation would allow the patient to avoid an ICD implant. WCD is best suited for clinical scenarios in which the risk of VT/VF is temporary or to bridge the patient to a more definitive treatment [4, 5]. To the best of our knowledge this is the first described use of WCD for a patient with suspected drug-induced LQTS, and, therefore, a temporarily elevated risk of VT/VF. We believe that LQTS differentiating protocol should include a WCD prescription throughout the whole drug washout period to avoid life-threatening arrhythmic events.



Article information

Conflict of interest: None declared.

Funding: None.

REFERENCES

1. Priori SG, Blomström-Lundqvist C, Mazzanti A, et al. 2015 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death: The Task Force for the Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death of the European Society of Cardiology (ESC). Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC). Eur Heart J. 2015; 36(41): 2793–2867, doi: 10.1093/eurheartj/ehv316, indexed in Pubmed: 26320108.
2. Wallace E, Howard L, Liu M, et al. Long QT syndrome: genetics and future perspective. Pediatr Cardiol. 2019; 40(7): 1419–1430, doi: 10.1007/s00246-019-02151-x, indexed in Pubmed: 31440766.
3. Sasaoka S, Matsui T, Hane Y, et al. Time-to-Onset analysis of drug-induced long QT syndrome based on a spontaneous reporting system for adverse drug events. PLoS One. 2016; 11(10): e0164309, doi: 10.1371/journal.pone.0164309, indexed in Pubmed: 27723808.
4. Sandhu U, Rajyaguru C, Cheung CC, et al. The wearable cardioverter-defibrillator vest: Indications and ongoing questions. Prog Cardiovasc Dis. 2019; 62(3): 256–264, doi: 10.1016/j.pcad.2019.05.005, indexed in Pubmed: 31077726.
5. Sterliński M, Oręziak A, Przybylski A, et al. Experts of the Heart Rhythm Section of the Polish Cardiac Society: opinion on the use of wearable cardioverter-defibrillators in Poland. Kardiol Pol. 2019; 77(2): 238–243, doi: 10.5603/KP.2019.0031, indexed in Pubmed: 30816992.

Figure 1. A 12-lead Holter electrocardiographic (ECG) monitoring strip showing an episode of nonsustained duomorphic ventricular tachycardia (**A**) and duomorphic ventricular extrabeats of the same morphology with a very short coupling interval superimposed on the T wave of a preceding beat (the "R-on-T" phenomenon) (**B**). A 12-lead ECG registered at 50 mm/s paper sweep speed presenting prolonged to 540 ms QT interval along with bifid T-waves that are asymmetrical and of low amplitude (**C, D**). The rhythm strip recorded by a wearable cardioverter-defibrillator vest showing ventricular fibrillation cardioverted with a 150-J biphasic shock delivery, resulting in restoration of sinus rhythm (**E**)