Successful Use of Quinidine in Treatment of Electrical Storm in Brugada Syndrome

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MOK, N.-S., ET AL.: Successful Use of Quinidine in Treatment of Electrical Storm in Brugada Syndrome. We report an adolescent with a malignant form of Brugada syndrome who presented with 15 episodes of ventricular fibrillation (VF) over 10 days, shortly after implantation of an implantable cardioverter defibrillator. Oral quinidine bisulphate at a dose of 1000mg/day successfully suppressed the electrical storm and recurrence of VF over 18-month follow-up. It also normalized the ST-segment elevation in his right precordial leads, suppressed all ambient unifocal ventricular extrasystoles and induction of VF on programmed electrical stimulation. This case suggests that quinidine, by virtue of its blocking action on Ito, may be useful as adjunctive therapy in Brugada syndrome. (PACE 2004; 27[Pt. I]:821–823)

Brugada syndrome, electrical storm, quinidine

Introduction

The last decade witnessed a large body of medical literature on the clinical profile of Brugada syndrome after its first description in 1992.1 To date, implantable cardioverter defibrillator (ICD) is the only effective treatment to prevent sudden death in patients suffering from Brugada syndrome while pharmacological treatment has no proven prognostic benefits. In this report we detail an adolescent with Brugada syndrome who presented with an electrical storm of ventricular fibrillation (VF) in whom oral quinidine bisulphate successfully suppressed the VF storm and prevented recurrence of VF without causing any side effects.

Case Report

An 18-year-old boy with mild mental retardation but otherwise good past health suddenly collapsed at home in March 2002. Automatic external defibrillator in ambulance documented recurrent VFs (five times) and successfully defibrillated all VFs. ECG recorded during sinus rhythm showed ST-segment elevation in leads V1–V2 and prominent J waves in precordial leads (Fig. 1A). Echocardiogram showed no evidence of structural heart disease. Holter monitoring recorded frequent unifocal premature ventricular complexes (PVCs). Viral study for myocarditis was negative. Coronary angiograms and right ventricular (RV) angiogram were normal. On programmed electrical stimulation (PES), VF was inducible with three ventricular extrasystolms delivered at RV outflow tract (RVOT) at baseline. We established the diagnosis of Brugada syndrome and implanted an ICD in this patient. Five days later he presented again after receiving several ICD shocks due to recurrent VFs. Intravenous amiodarone failed to prevent VF recurrence. Intravenous isoprenaline transiently suppressed the VF storm but VF quickly recurred following its discontinuation. A total of 15 VFs were recorded and successfully defibrillated by the ICD with 15 J over a period of 10 days. All VF episodes were initiated by the same PVC (Fig. 1B) with a left bundle branch block (LBBB) pattern and inferior axis suggesting an RVOT origin. Radiofrequency catheter ablation targeting the initiating PVCs was suggested as an adjunctive treatment but was refused by the patient’s parents. Finally we treated the patient (body weight 40 kg) with quinidine bisulphate at a dose of 1000mg/day. Quinidine successfully suppressed the electrical storm. During the steady state treatment, the ST-segment elevation in his right precordial leads was normalized, all ambient unifocal PVCs were suppressed on repeated Holter monitoring, and VF was rendered noninducible on PES (Fig. 2). The drug was well tolerated by the patient. Serial ECGs did not find any prolongation of his QTc interval. ICD did not record any recurrence of VF over 18-month follow-up.

Discussion

Antzelevitch et al. elegantly showed that Ito current in RV epicardium plays a pivotal role in the pathogenesis of Brugada syndrome.2 The only agent on the market with significant Ito-blocking properties is quinidine. Experimental studies have shown that quinidine, by blocking Ito on RV epicardium, is effective in restoring the epicardial action potential dome and thus normalizing the ST-segment and preventing Phase 2 reentry and
polymorphic VT/VF in experimental models of Brugada syndrome. To date, successful use of quinidine in treatment of Brugada syndrome is limited only to a few anecdotal reports and a retrospective study but has never been proven in any randomized trial. Alings et al. reported the phenomenon of quinidine induced electrocardiographic normalization in 2 patients with Brugada syndrome. Suzuki et al. reported an infant case with a malignant form of Brugada syndrome in which combined oral treatment with quinidine, denopamine, and atropine, ameliorated the ST-segment elevation in leads V1–V2 and suppressed recurrences of VT/VF for 6 months. Belhassen et al. in a retrospective analysis found that in all four patients with Brugada syndrome in whom quinidine (1000–2000 mg/day) prevented induction of VF, quinidine successfully prevented sudden death over a mean follow-up of 80.5 months. In our patient, quinidine given at a dose of 1000 mg/day has convincingly demonstrated its therapeutic effects in Brugada syndrome by suppressing the electrical storm of VF which was refractory to intravenous amiodarone, normalizing the ST-segment elevation in leads V1–V2, suppressing all ambient unifocal PVCs which were shown to initiate all the VF episodes, preventing the induction of VF on PES and finally, preventing recurrence of VF over 18-month follow-up without causing any side effects. Thus our case report added additional information to medical literature on the potential therapeutic value of quinidine in Brugada syndrome. However, its efficacy and long-term safety in the treatment of Brugada syndrome remain to be proven in large randomized trials.

Figure 1. (A) ECG (V1–V6) recorded during sinus rhythm showing ST-segment elevation in leads V1–V2 and prominent J waves in leads V3 and V4. (B) ICD electrograms showing three representative VF episodes. Note that the initiating PVCs (#) and the preceding PVCs (*) in all episodes carry the same morphology.
Figure 2. *Therapeutic effects of steady-state quinidine treatment in our patient with Brugada syndrome. (A) Normalization of ST-segment elevation in leads V₁–V₂ without prolongation of QTc interval. Note that J wave in lead V₃ also disappeared. (B) Previously inducible VF rendered noninducible during programmed electrical stimulation of RV apex performed through the ICD lead.*

References