Available online at www.sciencedirect.com
ScienceDirect
Journal of Electrocardiology 41 (2008) 325–328
JOURNAL OF Electrocardiology
www.jecgonline.com

Oral verapamil effectively suppressed complex ventricular arrhythmias and unmasked U waves in a patient with Andersen-Tawil syndrome

Okan Erdogan, MD,[a,*] Alper Aksoy, MD,[b] Nilda Turgut, MD,[b] Elcim Durusoy, MD,[b] Murat Samsa, MD,[b] Armagan Altun, MD[b]

[a]*Department of Cardiology, School of Medicine, Marmara University, İstanbul, Turkey*
[b]*Department of Cardiology, School of Medicine, Trakya University, Edirne, Turkey*

Received 29 September 2007

Abstract Andersen-Tawil syndrome (ATS) is a rare, heterogeneous, autosomal dominant, or sporadic disorder characterized by the clinical triad of periodic paralysis, dysmorphic features, and ventricular arrhythmias such as bidirectional ventricular tachycardia (BVT). We present a case of an elderly patient with ATS whose symptomatic ventricular arrhythmias including BVT were effectively suppressed by oral verapamil therapy.

Keywords: Andersen-Tawil; Verapamil; Bidirectional VT; U wave; Long QT; Therapy; Sudden death

Introduction

Andersen-Tawil syndrome (ATS) is a rare, heterogeneous, autosomal dominant, or sporadic disorder characterized by the clinical triad of periodic paralysis, dysmorphic features, and ventricular arrhythmias.[1] One gene, *KCNJ2*, has been identified so far and more than 20 mutations have been reported. Reduced IK1 resulting from *KCNJ2* mutations alters late cardiac repolarization by increasing its duration and leads to both distinctive T-U wave morphology and an increased propensity to ventricular arrhythmias.[2] Although the frequency and pattern of ventricular arrhythmias vary considerably among affected subjects, it is a common finding and includes frequent premature ventricular complexes (PVCs), bigeminy, polymorphic ventricular tachycardia (PVT), and bidirectional VT (BVT). Despite frequent VT burden, most ATS patients are remarkably asymptomatic, and sudden cardiac death is exceedingly rare.[1-8] However, management of complex ventricular arrhythmias and prevention of cardiac arrest still remain elusive given the lack of large comparative clinical studies. We present here a case of an elderly lady who incidentally was diagnosed with ATS and successfully treated with oral verapamil for symptomatic complex ventricular arrhythmias including BVT.

Case presentation

A 65-year-old woman with a history of hypertension and abdominal surgery for hydatid cyst in the liver presented to the emergency department with presyncope, palpitation, and generalized muscle weakness associated with nausea and vomiting. Surface electrocardiogram (ECG) demonstrated that the rhythm was nonsustained VT (12 beats) consisting of polymorphic PVCs that originate late after T wave from prominent U waves and a short run of BVT preceded by sinus complex (Fig. 1). During previous admissions for symptomatic ventricular arrhythmias, both β-blocker and propafenone as well as sotalol, whose first dose (40 mg) once elicited torsades de pointes according to past medical records, were ineffective for suppression of arrhythmias. Family history revealed that her brother suddenly died of unknown cause at a young age. Coronary angiography and transthoracic echocardiography performed during last admission showed normal findings. Her physical examination revealed short stature (height, 1 m 40 cm; weight, 40 kg), micrognathia, small hands and feet, and clinodactyly of fifth fingers in both hands. Her blood potassium level on admission was 3.5 mmol/L, although successive values on subsequent days showed slight variations (range, 3.8-4.7 mmol/L). Neurological evaluation revealed that she was having recurrent episodes of mild proximal muscle weakness beginning from childhood. She also referred difficulty releasing her hand grip. Her global cognitive status was impaired. Standardized mini-mental test (SMMT-E)

* Corresponding author. Marmara Üniversitesi Tip Fakültesi Hastanesi, Altunizade, İstanbul, Turkey. Tel.: +90 216 327 10 10; fax: +90 212 327 60 35.
E-mail address: okanerdogan@yahoo.com

0022-0736/$ – see front matter
doi:10.1016/j.jelectrocard.2008.02.023

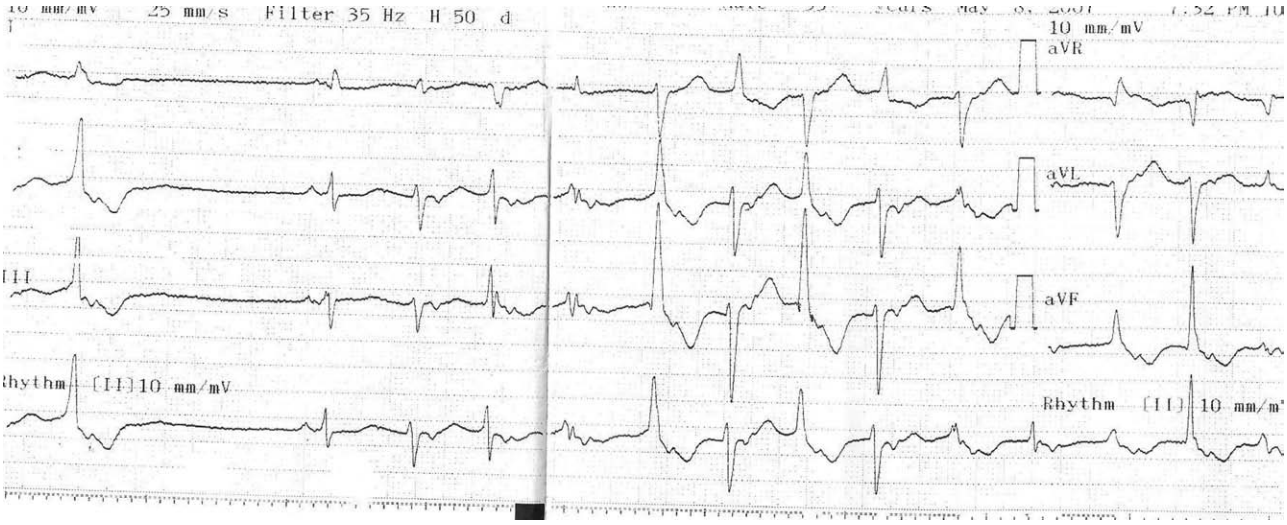

Fig. 1. Nonsustained VT episode (12 beats) consisting of polymorphic PVCs and a short run of BVT preceded by sinus complex.

score was calculated as 15 (normal, >24). Suggesting intracellular calcium overload and triggered activity as the probable mechanisms, we speculated that empiric therapy with verapamil might be effective in suppressing frequent BVT episodes. On the third day of oral verapamil (40 mg TID) therapy, BVTs were suppressed before the rhythm initially converted to bigeminy and then to sinus rhythm with interspersed PVCs every other third or fourth beat. We were then able to reliably measure the baseline ECG intervals (Fig. 2). The PR interval and QRS duration were normal. On precordial leads, there were prominent U waves maximally measured in amplitude as 0.3 mV, with a maximal duration of 200 milliseconds in lead V5. The interval between T peak and U peak was significantly prolonged and measured 240 milliseconds. Both QU and QUc intervals were prolonged and maximally measured 720 milliseconds, whereas QT and QTc intervals remained normal and were measured as 440 milliseconds at a heart rate of 60 beats per minute, respectively. T-wave amplitude and duration were 0.15 mV and 120 milliseconds, respectively. The QU dispersion measured in all leads including limb leads was calculated to be 60 milliseconds, which was mainly related to

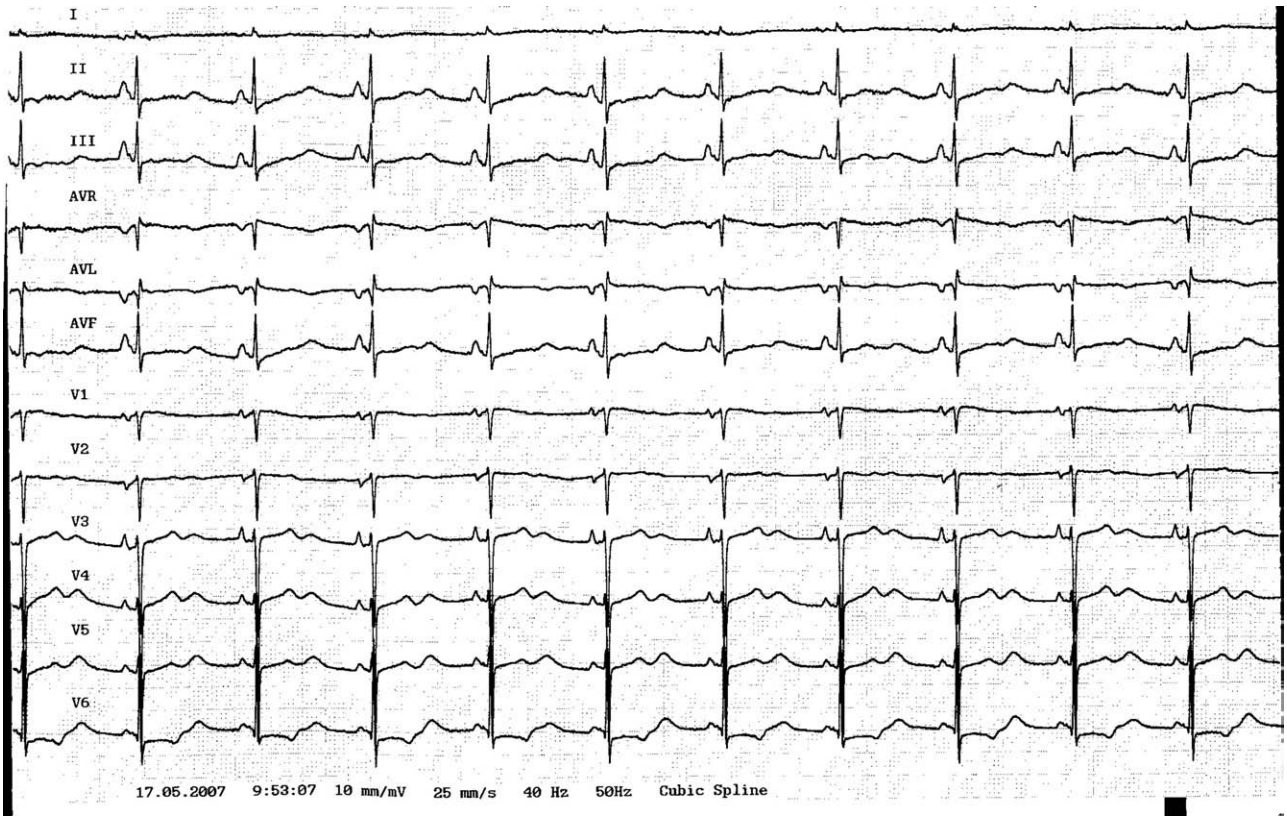

Fig. 2. Surface ECG was recorded after verapamil therapy was started, and complex ventricular arrhythmias were suppressed.