

# A case of brugada syndrome presenting with ventricular fibrillation storm and prominent early repolarization<sup>☆</sup>

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## Abstract

A 21-year-old man developed ventricular fibrillation (VF) while drinking alcohol and was admitted to our hospital. An electrocardiogram (ECG) on admission revealed remarkably prominent slurs on the terminal part of QRS complexes in the left precordial leads and a coved type ST elevation at higher intercostal spaces. After hypothermia therapy, he underwent implantation of an implantable cardioverter-defibrillator (ICD). Standard twelve-lead follow-up ECGs revealed early repolarization pattern and an intermittent coved type ST elevation. When the coved type ST elevation appeared, the early repolarization pattern in the inferior and left precordial leads was attenuated. Prominent early repolarization pattern was the most likely trigger of the VF storm in this Brugada patient.

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## Keywords:

Brugada syndrome; Idiopathic ventricular fibrillation; VF storm; Early repolarization syndrome

## Introduction

Brugada syndrome (BrS) is an inheritable channelopathy characterized by coved-type ST elevations on the right precordial leads and is associated with a potential risk of sudden death due to ventricular fibrillation (VF) [1]. Moreover, early repolarization pattern, slur or notch ( $\geq 0.1$  mV in amplitude) in the terminal part of the QRS complex can be another hallmark of non-Brugada-type idiopathic VF patients [2]. The combination of BrS and early repolarization syndrome is not rare and has been considered to be a risk factor for the occurrence of VF [3,4].

We report a young male patient with BrS combined with early repolarization syndrome who was hospitalized for recurrent VF. Of the two arrhythmogenic substrates, the prominent early repolarization pattern were considered to be the trigger of the VF storm.

## Case

A 21-year-old male developed syncope for the first time after heavy drinking at night and was transferred to another hospital 2 weeks prior to admission to our hospital. Because the physical examination did not disclose abnormalities, and his ECG was considered normal, he was allowed to go home. Subsequently, he lost consciousness at approximately 3:20 a.m. while drinking with friends. A VF was documented and the patient was defibrillated by emergency personnel using an automatic external defibrillator (AED). On route to our hospital, the VF recurred, and the patient was defibrillated. He was admitted to our hospital at 3:34 a.m. on the 28th of June.

His past history was non-contributory with the exception of an arrhythmia that was noted when he was a senior high school student. He was a heavy drinker and a heavy smoker. He had no family history of sudden cardiac death or BrS.

## Examinations on admission

The patient was unresponsive, but his pupils responded to light. His blood pressure was 100/70 mmHg (systolic/diastolic), and he exhibited sinus tachycardia (127 beats/min). His body temperature was 37.5 °C. Otherwise, the physical examinations were normal. There were no abnormalities in blood chemistry, complete blood counts, or serum electrolytes. Later, his body

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length and weight were measured as 172 cm and 65 kg, respectively.

Echocardiography revealed normal chambers with normal motions and no pericardial effusion. Computed tomography was negative for pulmonary embolism and aortic dissection. Emergency catheterization revealed normal coronary arteries and normal cardiac function. After the examinations, he was placed under therapeutic hypothermia at 34 °C with assisted respiration.

### Course in the hospital

His ECG on admission revealed sinus tachycardia and slurs on the descending limb of the R in leads V3–V5 that were considered to represent markedly prominent early repolarization pattern of 0.3–0.6 mV in amplitude. Early repolarization pattern of 0.2 mV in a VF were present in the inferior leads; they were <0.1 mV in leads II and III (Fig. 1). The prominent early repolarization pattern disappeared soon after admission.

VF developed again at the time of intubation, and the patient was defibrillated. Under hypothermia therapy, amiodarone treatment was initiated but was soon terminated due to the prolongation of the QTc to 518 ms<sup>1/2</sup> (Fig. 2). The early repolarization pattern reappeared both in the inferior and left precordial leads, and thereafter, early repolarization pattern, were persistently observed in the inferior lead but were absent or present at only trace levels in the left precordial leads (Fig. 2). Because the ST morphologies in V1 and V2 were variable, ECGs were recorded from higher intercostal spaces, and a diagnosis of BrS was made (Fig. 2). A coved-type ST elevation was constantly observed at the

higher intercostal spaces when ECGs were intermittently collected.

On the third day of the hypothermia therapy, he regained consciousness, and he was warmed and extubated. Mild disturbances in memory, concentration and calculation function were observed. One month later, he was implanted with an implantable cardioverter-defibrillator (ICD). After reprogramming the ICD due to inappropriate shocks for sinus tachycardia, he was discharged on the 29th day of hospitalization.

### Follow-up

One month after discharge, the patient received an ICD shock during a rehabilitation program, and he was readmitted. The ICD memory revealed an appropriate shock for VF. Bepridil (100 mg/day) was then added to prevent VF, and the patient was discharged and followed in the outpatient clinic of our hospital. He has been asymptomatic for 1.5 years.

Recent ECGs revealed a fully paced atrial rhythm of 72 beat/min with normal AV conduction. The ECGs also revealed occasional coved-type ST elevations in V1–2 (Fig. 3) and slurs or notches in the inferior and left lateral leads. Upon the appearance of the coved-type ST elevations, the early repolarization patterns were apparently attenuated (Fig. 3). The genetic screening was not performed.

### Discussion

A 21-year-old male with BrS was rescued from sudden death due to recurrent VF. The main findings are as follows:

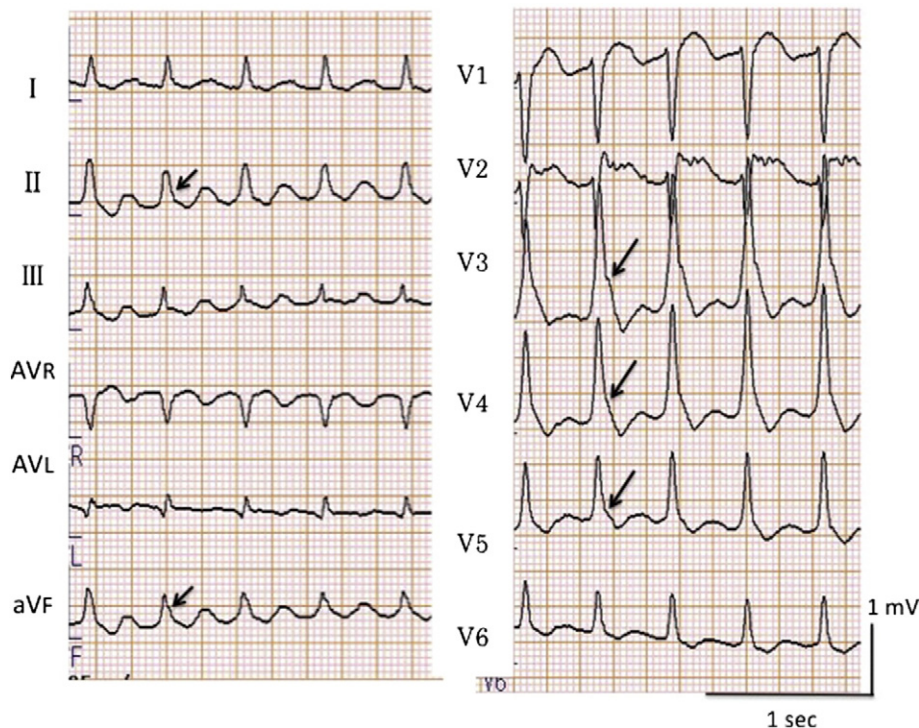


Fig. 1. ECGs on admission. ECG on admission approximately twenty min after defibrillation showing sinus tachycardia at a rate of 127 beats/min. In V3 and V5, a steeply declining ST-segment towards the T wave indicative of prominent early repolarization (arrows) was observed. V1 and V2 revealed a non-specific elevation of the ST-segment.

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