



CASE REPORT

Syncopal monomorphic ventricular tachycardia with pleomorphism, sensitive to antitachycardia pacing in a patient with Brugada syndrome

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Polymorphic ventricular tachycardia and ventricular fibrillation are the most common arrhythmias in Brugada syndrome, causing syncope or sudden death. Sustained monomorphic ventricular tachycardias are rare in this context. We report the case of a 41-year-old man with repetitive syncopal episodes and an ajmaline-induced characteristic Brugada ECG pattern, in whom episodes of monomorphic ventricular tachycardia with pleomorphism and response to ventricular pacing were documented.

Case report

A 41-year-old man was referred to our Arrhythmia unit after a history of recurrent syncope and pre-syncope during the past 14 months. The patient had no family history of syncope or sudden death. The findings of the initial evaluation (including clinical history, physical examination, baseline ECG, carotid sinus massage, postural blood pressure testing, 24 h ambulatory ECG monitoring, echocardiogram, and cardiac magnetic resonance imaging) did not disclose the cause of syncope. A head-up tilt test was negative. The patient experienced recurrent pre-syncope, some of the episodes clearly related to neurally mediated triggers, and sudden syncopal episodes with body injury. Tilt test with nitroglycerine challenge was repeated and remained normal. An electrophysiological study was indicated and performed in the post-absorptive non-sedated state. The basal 12-lead ECG at the time of the electrophysiological study (*Figure 1A*) showed sinus rhythm at 70 bpm, PR interval 140 ms, QRS duration 110 ms, and QT interval 400 ms. The QRS in leads V₁ and V₂ showed an rSr' pattern and doubtful (<1 mm) ST-segment elevation (Brugada ECG type 3). Tests of sinus and atrioventricular node function were within normal ranges. The HV interval was 50 ms and a sporadic infraHisian block of atrial premature beats was observed. Programmed right atrial and ventricular stimulation (increasing heart rates and up to three extrastimuli at two basic cycles, before and after isoprenaline infusion)

did not induce significant arrhythmias. After administration of 50 mg intravenous of ajmaline, the ECG showed a coved-type ST-elevation (Brugada ECG type 1) in leads V₁ and V₂ (*Figure 1B*).

Under the suspicion of syncope in the Brugada syndrome, an implantable loop recorder (ILR) (Reveal 9525, Medtronic Inc., Minneapolis, MN, USA) was implanted after the electrophysiological study, trying to document further the mechanism of syncope. In the first week after implantation, the patient experienced two syncopal episodes and activated the device. The stored electrograms were retrieved by telemetry, and in both activations, a self-sustained monomorphic regular ventricular tachycardia was detected, but the QRS polarity and morphology were different in each episode (*Figure 2*). A defibrillator was implanted, and during the follow-up, appropriate shocks were delivered by the device, always in response to monomorphic regular fast tachycardias. After these findings, antitachycardia pacing therapies were enabled (burst and ramp pacing), and episodes of monomorphic ventricular tachycardia of at least two morphologies have been detected and successfully treated with ventricular burst pacing (*Figure 3*).

Syncopal attacks in the Brugada syndrome are typically related to polymorphic ventricular tachycardia,¹ but cases of monomorphic ventricular tachycardia have also been reported in the literature.²⁻⁵ In this patient, episodes of monomorphic regular ventricular tachycardia, with pleomorphism and sensitive to ventricular stimulation, were the cause of syncopal episodes. These arrhythmias could not be induced during the electrophysiological study, but were detected during the follow-up with the use of an ILR.

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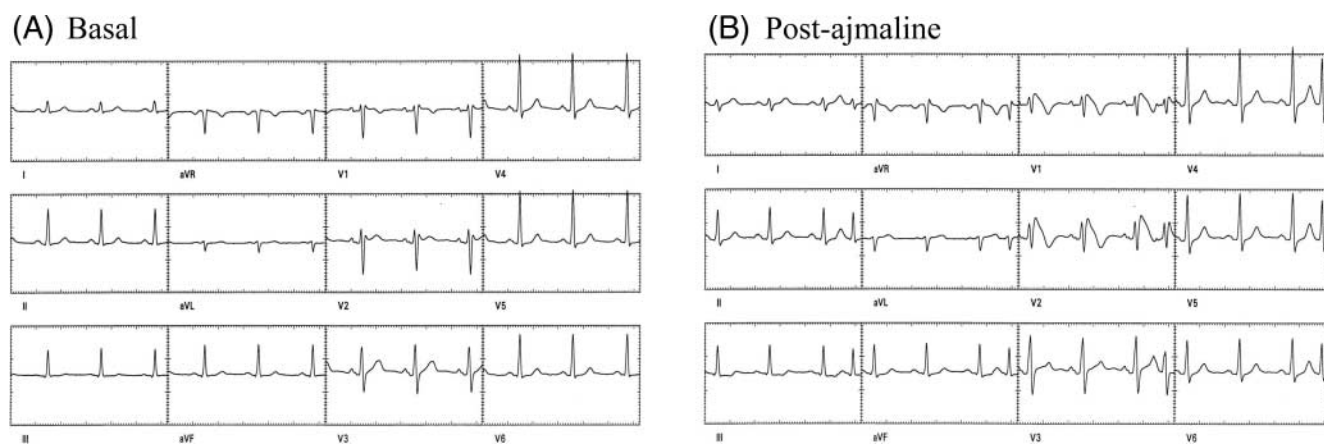


Figure 1 (A) Basal 12-lead ECG and (B) ECG post-ajmaline.

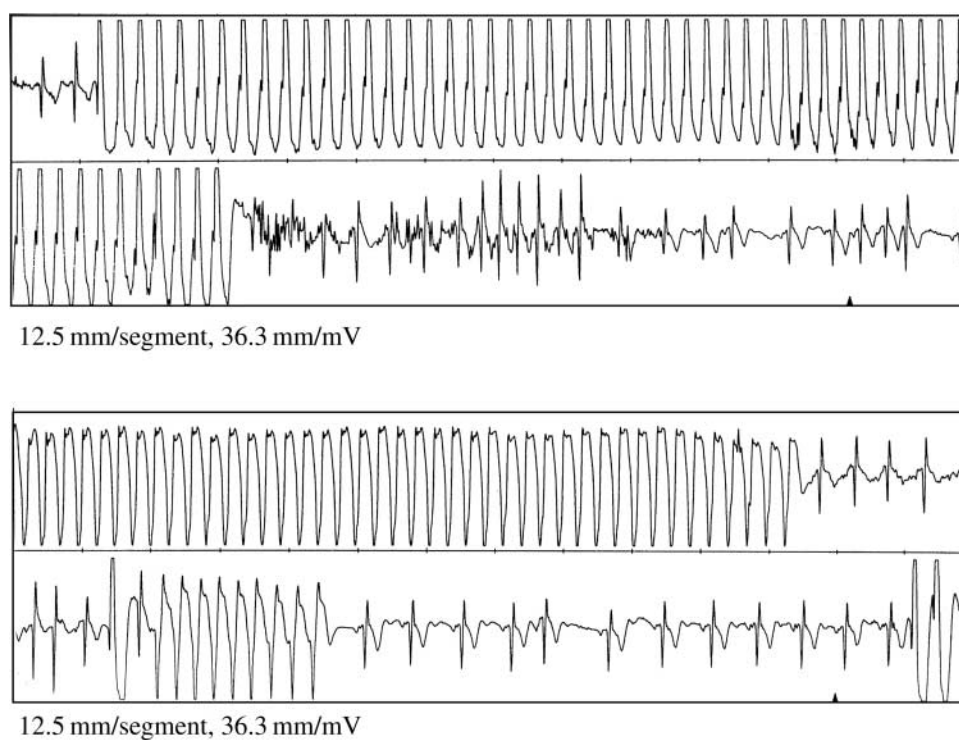


Figure 2 Implantable loop recording during two syncopal episodes.

The present case fulfils the Brugada syndrome diagnostic criteria because of the presence of syncopal episodes and a type 1 ECG after the administration of a sodium channel blocker.¹ In addition, no structural heart disease could be detected in the initial evaluation or in the follow-up. Two unique features in the present case are the detection of two distinct types of monomorphic ventricular tachycardias during the syncopal attacks and the fact that ventricular burst pacing has been effective in the suppression of those ventricular tachycardias. It is noteworthy that during electrophysiological study or follow-up, no polymorphic tachycardia has been registered.

A differential diagnosis must be performed considering Brugada syndrome, idiopathic fascicular tachycardias, and other types of ventricular tachycardias. The absence of

inducibility after isoprenaline infusion and the presence of two morphologies of the QRS make very unlikely the existence of other causes of tachycardia, although, as we have mentioned earlier, the patient fulfils the Brugada syndrome diagnostic criteria.

To the best of our knowledge, these findings have not been described previously in the Brugada syndrome. Although arrhythmias in the Brugada syndrome usually are not sensitive to ventricular pacing, in this case, stimulation was effective in the suppression of the tachycardia. These two features (monomorphic pattern and response to stimulation) would suggest a re-entrant mechanism.⁶

Dysfunction of the sodium channel seems to be the centre of the pathogenesis of the Brugada syndrome in most cases in response to a mutation in the gene SCN5A. Although

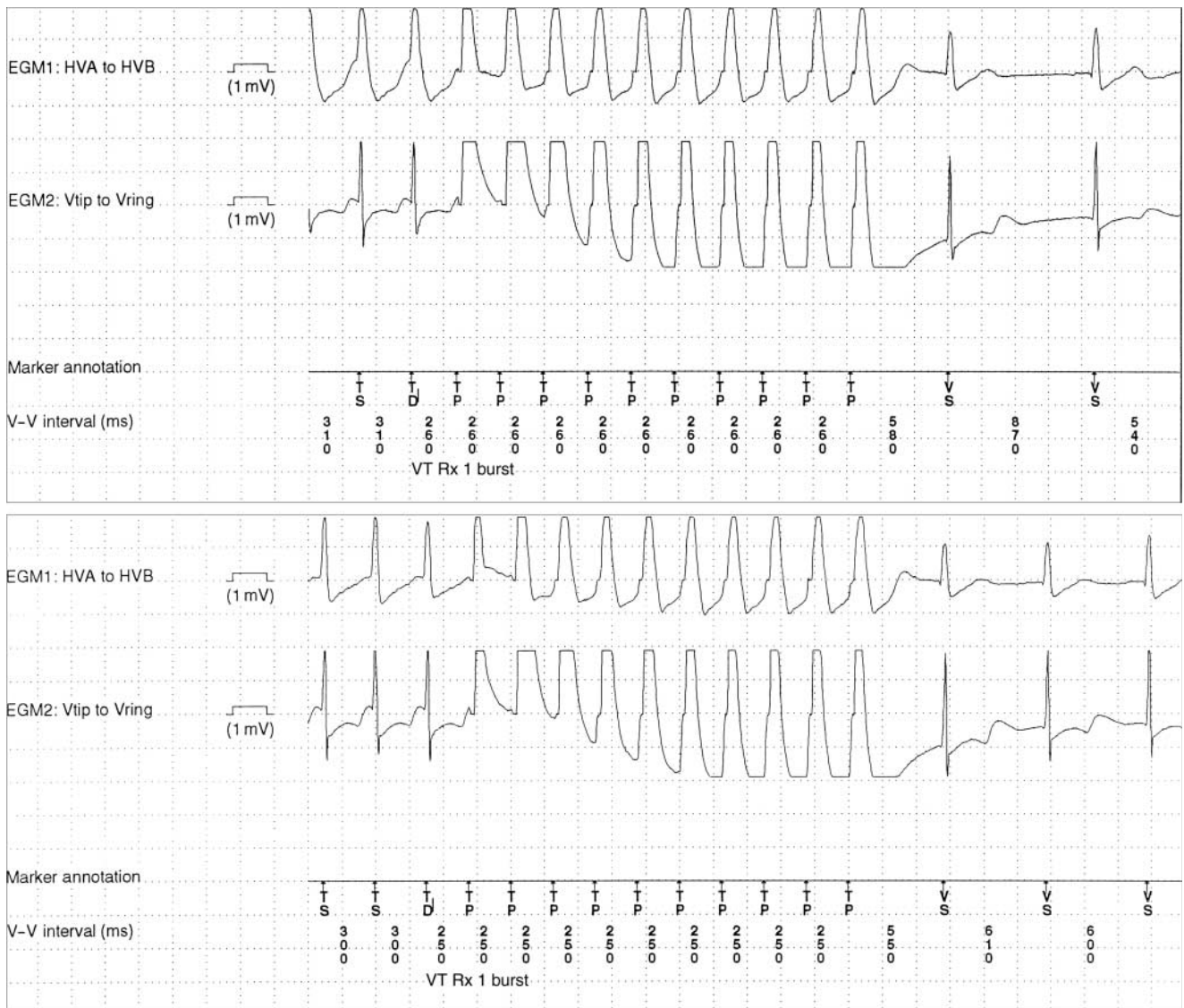


Figure 3 Electrograms of two syncopal episodes. The upper panel shows a non-sustained monomorphic ventricular tachycardia. The lower panel shows the sustained monomorphic ventricular tachycardia in response to ventricular burst pacing. Note that electrograms in both panels show different polarities and morphologies (pleomorphism).

SCN5A defect has been related to late potentials and other electrical phenomena in the right ventricle outflow tract, this channel is spread all over the myocardium. In fact, various electrophysiological disturbances have been described in the setting of the Brugada syndrome: higher incidence of atrial fibrillation (up to 10%) and other supra-ventricular arrhythmias (20%),⁷ prolonged sinus node recovery time and sinoatrial conduction time, PR and HV interval prolongation, or a slight prolongation of the QT interval.¹ The broad nature of these anomalies should explain the possibility of finding different clinical arrhythmias (monomorphic ventricular tachycardia, in our case) caused by a unique molecular mechanism.

Although Brugada syndrome is usually linked to polymorphic ventricular tachycardia, some atypical presentations can be found, including monomorphic, pleomorphic, and pacing-sensitive ventricular tachycardia.

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