

# Supraventricular Tachyarrhythmia and Brugada Syndrome: A Case of Atrial Fibrillation and Brugada Syndrome in a Young Patient Without Structural Heart Disease

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## ABSTRACT

*We report a case of a 29-year old man who initially presented with a single episode of syncope. The initial electrocardiogram (ECG) showed atrial fibrillation and an ST segment elevation on lead V1. A flecainide test unmasked the Brugada syndrome. The pathophysiology of Brugada syndrome and atrial fibrillation in this patient could be connected by sodium channel dysfunction throughout the heart. In addition, we reviewed the possible connection between Brugada syndrome and atrial fibrillation.*

**Keywords:** Atrial fibrillation, Brugada Syndrome, channelopathy

## Taquiarritmia Supraventricular y Síndrome de Brugada: un caso de Fibrilación Atrial y Síndrome de Brugada en un Paciente Joven sin Cardiopatía Estructural

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## RESUMEN

*Reportamos el caso de un hombre de 29 años de edad que se presentó inicialmente con un solo episodio de síncope. El electrocardiograma inicial (ECG) mostró fibrilación atrial y una elevación del segmento ST en la derivación V1. Una prueba de flecainida reveló la presencia del síndrome de Brugada. La patofisiología del síndrome de Brugada y la fibrilación atrial en este paciente podrían estar conectados por una disfunción del canal de sodio a través del corazón. Además, examinamos la posible conexión entre el síndrome de Brugada y la fibrilación atrial.*

**Palabras claves:** Fibrilación atrial, síndrome de Brugada, dysfunction del canal de sodio

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## INTRODUCTION

Brugada Syndrome (BS) is an inherited cardiac disorder initially described in 1992 by Pedro and Josep Brugada (1) with variable electrocardiographic features characteristic of right bundle branch block, persistent ST segment elevation in the precordial leads (V1–V3) at rest and sudden cardiac death.

Brugada Syndrome presents in a certain number of patients as an inherited cardiac arrhythmic disorder caused

by mutations in the cardiac sodium channel gene *SCN5A* (2). It manifests mainly in adults with sudden death peaking around 40 years of age (3).

We report the case of a 29-year old man with no recorded history of heart disease who initially presented with a single episode of syncope and atrial fibrillation (AF) with an abnormal ST segment elevation in lead V1.

## CASE DESCRIPTION

A 29-year old man, with no history of heart disease or any cardiovascular risk factors, presented to the emergency room of our centre because of a single episode of syncope accompanied by tonic-clonic seizures and sphincter disturbances. The patient briefly lost consciousness during the episode and awoke on his own after a brief period without resuscitative manoeuvres. In the initial presentation, the patient was haemodynamically stable and the electrocardiogram (ECG),

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showed atrial fibrillation at 150 bpm. The ECG tracing also showed a mild convex ST segment elevation on V1 with no significant modifications in the duration of the QRS. The patient was rate controlled with a calcium channel blocker (diltiazem) and spontaneously converted to sinus rhythm and was discharged home with electrophysiologic (EP) follow-up and asked to return for monitoring 48 hours after the initial episode.

At the next visit, the patient presented in sinus rhythm with a persistent ST segment elevation on lead V1. While the initial diagnosis was syncope of probable vasovagal origin but when the image on V1 persisted, the flecainide challenge test (150 mg/10 minutes) was performed which unmasked BS (Fig. 1) and a significant widening of the QRS on V1 and

the case of the index patient who was initially diagnosed with AF, with BS being diagnosed subsequently.

The diagnosis in the patient was AF associated with BS, and we suggest that the patient had an episode of VF and briefly lost consciousness. He was in AF instead of sinus rhythm after the episode, as exemplified by the initial ECG, which showed AF at 150 bpm. Additionally, a mild convex ST segment elevation on lead V1 was observed which persisted at the next visit and was the reason for performing the flecainide challenge test which unmasked the BS.

Brugada Syndrome should be included in the differential diagnosis of any patient with syncope, especially young patients, and when the electrocardiogram indicates a Brugada pattern or when risk factors such as a family history

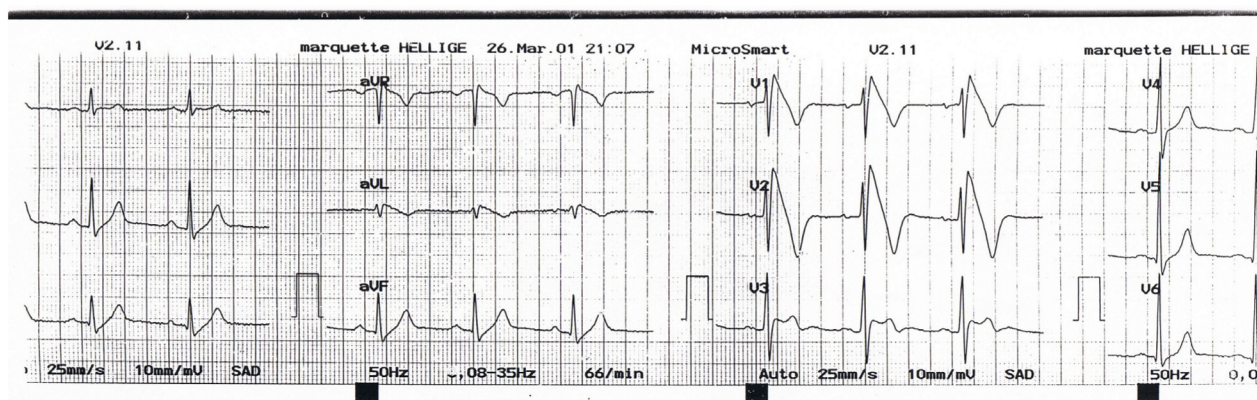


Fig. 1: ECG showing ST-segment elevation on precordial leads (V1–V2).

V2 with convex rapid descent elevation of the ST segment of 3 mm in these leads. During the last minutes of the test, a polymorphic ventricular tachycardia (VT) was observed associated with a feeling of instability and passing out. It was concluded that the electrophysiologic study (EPS) triggered right ventricular tachycardia with scaled stimulation: therefore the patient was scheduled for an automated implantable defibrillator device placement.

## DISCUSSION

We report the case of a 29-year old man with no history of heart disease or any cardiovascular risk factor who presented with syncope. Identification of the underlying cause of syncope is difficult and is not established in up to 18% of cases (4). However, the diagnosis, which can vary from benign cases of vasovagal syncope to heart disease related syncope is associated with a more than twofold increase in mortality (5).

Although cardiac-related causes account for relatively a small proportion of cases of syncope (14%), they should always be excluded due to the worse prognosis (6). The ECG is an important part of the initial assessment of any patient with syncope, but is diagnostic in only 5% of cases and suggests a potential cause in another 5% of patients (7), as is

of sudden death exists.

The arrhythmogenic substrate in BS is not necessarily restricted to the right ventricle (7); the association of BS with AF or supraventricular tachycardias is rare although some cases have been reported (8–11).

Eckard *et al* (8) found that supraventricular tachyarrhythmia was induced by programmed atrial stimulation in 10 of 35 patients with BS (AF 1/10). They concluded that the arrhythmogenic substrate in BS may not be restricted to the ventricular level and that BS should be considered as possible additional electrophysiologic abnormality in patients with supraventricular tachyarrhythmia presenting with sudden cardiac death or syncope.

Itoh *et al* (9) retrospectively analysed 30 patients with a Brugada-type electrocardiogram (Brugada-ECG) and found that paroxysmal AF was present in 9 out of 30 patients (30%) with BS, suggesting that a Brugada-ECG may be associated with an increased risk of both ventricular tachyarrhythmias and paroxysmal AF and that the arrhythmogenesis may be related to the pronounced ST-segment elevation.

Morita *et al* (10) compared 18 patients with BS with a group with no organic heart disease and concluded that atrial vulnerability is increased in patients with BS. They found that spontaneous AF occurred in 7 of the 18 (39%) patients

with BS but not in the control group and that AF was induced by programmed atrial stimulation in 8 of 14 patients (57%) with BS.

The right atrial effective refractory period (RA-ERP) did not differ between the two groups. The intra-atrial conduction time was increased in the BS group compared with the control group ( $168.4 \pm 17.5$  vs  $131.8 \pm 13.0$  ms,  $p < 0.001$ ). The duration of atrial potential in the RA-ERP was prolonged in the BS group versus the control group ( $80.3 \pm 18.0$  vs  $59.3 \pm 9.2$  ms,  $p < 0.001$ ). Repetitive atrial firing was induced in 9 patients with BS and in 6 control subjects. Atrial fibrillation was induced in 8 patients with BS but in none in the control group. In patients with BS without spontaneous AF, the intra-atrial conduction time and duration of atrial potential were also increased.

Bordachar *et al* (11) in a single-centre prospective study of 59 consecutive patients with BS found that the incidence of atrial arrhythmias in patients with a spontaneous electrocardiogram with BS was 26% vs 10% in patients with a flecainide-induced electrocardiogram ( $p < 0.05$ ). In patients with an indication for an implantable cardioverter defibrillator (ICD), the incidence of atrial arrhythmias reached 27% vs 13% in patients with BS but with no indication for an ICD ( $p < 0.05$ ); patients with BS had an abnormally high proportion of atrial arrhythmias. Their results argue against a structural form of atrial disease, but suggest a pure electrical aetiology of atrial arrhythmias as an extension of the arrhythmogenic substrate of BS to the atrial level.

Recently Yamada *et al* (12) investigated whether BS was associated with vulnerability not only to ventricular fibrillation but also to atrial fibrillation, in 15 patients with BS and Brugada-type electrocardiogram and concluded that the electrical abnormality in BS is not limited to the ventricular level as similar changes occur in the atria. Such abnormal conduction properties could be a substrate for re-entrant atrial tachyarrhythmias.

A genetic correlation between BS, long QT syndrome, and sinus node dysfunction has been proposed (13–17). The genetic abnormalities causing BS have been linked to mutations in the SCN5A gene on chromosome 3p21–23, encoding for the cardiac sodium channel (14). Mutations in the cardiac sodium channel  $\alpha$  subunit gene NaV 1.5 have been associated with a spectrum of clinical rhythm disorders, including long QT syndrome type 3 (LQT3), BS and progressive cardiac conduction system disease [PCCD] (15). If a mutation in the cardiac sodium channel does in fact cause BS (18–20), a myocardial electrical abnormality might exist not only in the ventricular myocyte but also in the atria (10). Sumiyoshi *et al* (20) studied 5 patients with symptomatic BS, and found that 4/5 had spontaneous episodes of ventricular fibrillation (VF), 1/5 had syncope and 3/5 had a documented sinus pause  $>3$  s (a 42-year old man, a 62-year old man and a 49-year old woman). Only one of them had a family his-

tory of sudden death; two also had atrial fibrillation or flutter. Electrophysiology demonstrated prolonged sinus node recovery time in two patients (2.6 s and  $>5$  s), fitted with a cardiac pacemaker after episodes of VF before the diagnosis of BS was made. They suggested that prolongation of the action potential of sinus node cells and slowing of diastolic depolarization due to an abnormal sodium channel gene may contribute to sinus node dysfunction. A reduced sodium current could account for BS and a conduction disturbance in the sinoatrial region (20), with slow atrioventricular conduction resulting in a slow ventricular response during atrial fibrillation.

It is believed that vagal activity plays an important role in ST-segment elevation and ventricular fibrillation in patients with BS (21) and this could be related to the initiation of paroxysmal AF and slower atrioventricular conduction in these patients. Atrial vulnerability is enhanced in BS patients with and without AF. Therefore, an electrical abnormality in the atrium exists in patients with BS who do not experience an attack of atrial fibrillation (7).

Finally, some studies suggest that patients with BS paroxysmal atrial arrhythmias may present with more advanced disease (11, 22) and patients who presented with a BS-type ECG only after administration of flecainide are thought to have a better prognosis (11), as in this patient.

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