

## Case Reports

# Brugada Syndrome in A Jordanian Patient

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### Abstract

Brugada syndrome is a new clinical entity first described by Brugada and Brugada in 1992. It has a characteristic ECG pattern but it can be missed. We report on a Jordanian patient who has been presented with this syndrome.

### Introduction

Brugada syndrome is one of several conditions that can cause sudden cardiac death in a patient with a structurally normal heart. Diagnosis is based on demonstration of the characteristic electrocardiographic patterns, caused by a mutation in the cardiac sodium channels.<sup>1</sup>

**Keywords:** Brugada Syndrome, Jordanian Patient.

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### Case

A 21-year-old male university student, previously healthy, referred to our hospital as a case of anteroseptal myocardial infarction. The history dates back to three days, when he felt non-specific pricking, localized to the left sub mammary chest pain. This is not associated with dyspnea, nausea, palpitation, and vomiting. The electrocardiogram showed sinus rhythm and ST-segment elevation in the precordial leads V1-V3 (figure1). The patient was diagnosed as anteroseptal myocardial infarction and was given streptokinase 1.5 million unit. Repeated electrocardiogram revealed persistent ST-segment elevation. He was transferred to our hospital as failed thrombolytic therapy for rescue angioplasty. Upon his arrival, he was asymptomatic with stable vital sign.

The electrocardiogram showed sinus rhythm, incomplete right bundle branch block pattern, and coving type ST-segment elevation in precordial leads V1,V2, and V3, which is characteristic of Brugada syndrome type 1 (figure 1). Laboratory workup revealed normal Complete Blood Count, Urea and Electrolyte, Calcium, Liver Function Test, Lipid profile, blood glucose, and negative Troponin. Cardiac catheterization showed normal coronary arteries and left ventricle. The patient and family reassured that he had no myocardial infarction.

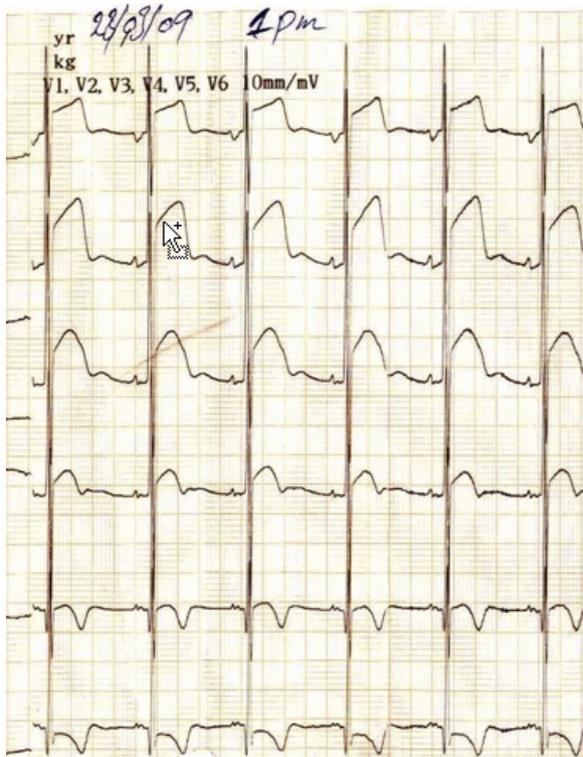
There was no family history of sudden cardiac death, syncopal attacks, or cardiac diseases. Family screening revealed abnormal electrocardiogram of his father (65 years old) and one brother (32 years old) (figure 2, 3) and normal electrocardiogram in the 2<sup>nd</sup> brother (28 years old). The patient was advised to have electrophysiological study.

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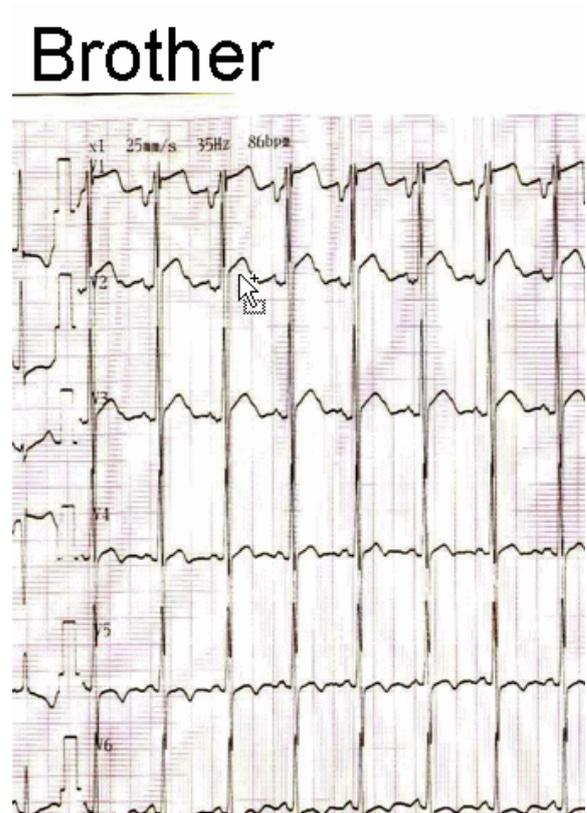
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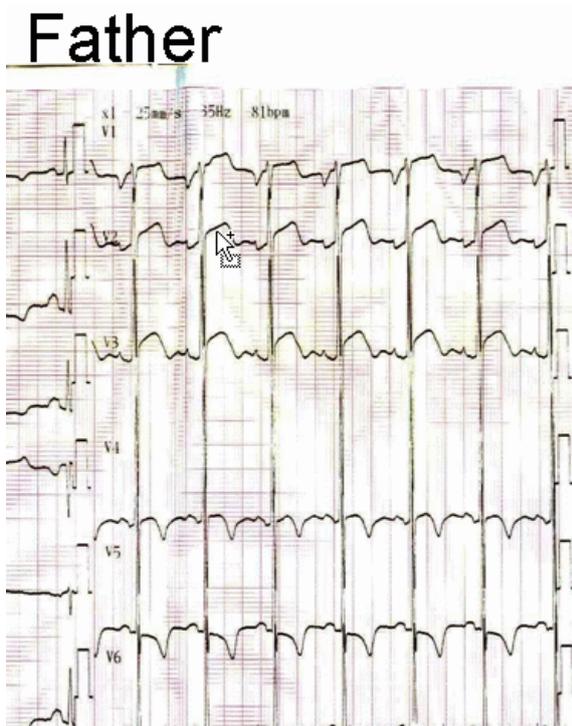
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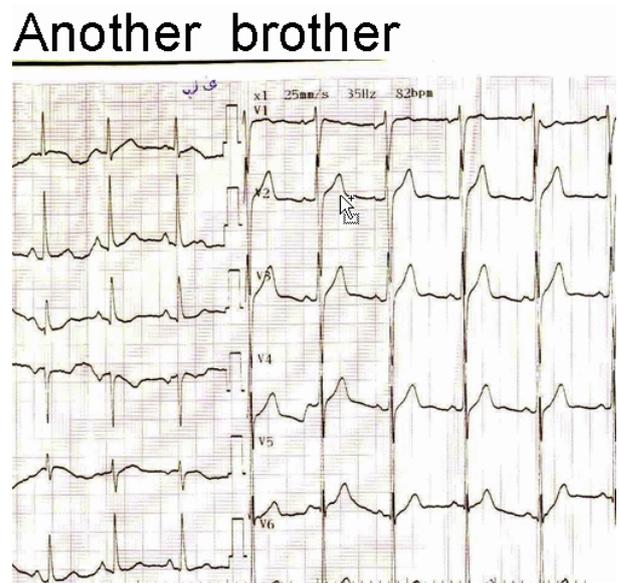
**Figure (1):** The Characteristic ECG of Brugada syndrome. ST-Segment elevation in leads V1-V3.



**Figure (3):** ST-Segment elevation in leads V1-V3.



**Figure (2):** The characteristic ECG of Brugada syndrome.



**Figure (4):** Normal ECG.

## Discussion

Brugada syndrome is a type of cardiac membrane channelopathy. It was first identified in 1992 by Pedro and Josep Brugada.<sup>1</sup> It is inherited through familial autosomal dominant transmission. A mutation, thought to be found on the SCN5A gene, leads to a decreased sodium inflow current, thereby reducing the duration of the normal action potential and resulting in a less opposed and more prominent outward potassium current.<sup>2, 3</sup> The net effect is a reduction in the duration of the normal action potential in the right ventricular epicardium, but not in the endocardium. The change in current is responsible for the characteristic ECG abnormalities seen in patient with Brugada syndrome and the tendency toward “phase-2 reentry” of the action potential, which can trigger ventricular arrhythmias. Consequently, a shorter-than-normal epicardial action potential occurs, which can lead to ST-segment elevation in the right precordial leads and to ventricular tachyarrhythmias.<sup>4</sup>

Brugada syndrome can occur in different population but is most common in Southeast Asian men. In one study, up to 0.7% of the 14,000 Japanese participants had a brugada-type ECG.<sup>5</sup> In one US teaching hospital, 52 (0.4%) of 12,000 noncardiac patients were found to have an ECG pattern consistent with brugada syndrome. However, the overall prevalence among normal population is not established because the ECG is dynamic and there are concealed forms of this syndrome.<sup>6</sup>

About 60% of patients with Brugada syndrome have a family history of sudden death or have family members with the ECG pattern of Brugada syndrome. Forty percent of patients have sporadic de novo mutation.<sup>7</sup>

Patient with Brugada syndrome may be asymptomatic and diagnosed accidentally (as in our patient), or presented with cardiac events. Most of the cardiac events due to ventricular fibrillation occur at night as a form of sudden cardiac death, agonal respiration, and syncope. It

is usually manifested during adulthood with a male predominance (male: female ratio, 8:1). All clinical manifestations of Brugada syndrome are attributed to ventricular tachyarrhythmias and their complications. Mostly, these arrhythmias occur at rest or during night time.<sup>8</sup> These can be explained by increased vagal tone.<sup>9</sup> Patients are usually presented in the third or fourth decade of life; just fewer than 50% of patients have a family history of sudden cardiac death. In addition to ventricular arrhythmias, patients are also at great risk of atrial arrhythmias. In a study of 59 patients with brugada syndrome and 31 controls, spontaneous atrial fibrillation occurred in 20% of the Brugada patients and none of the controls.<sup>10</sup>

The Brugada syndrome is associated with a peculiar pattern on the ECG consisting of a pseudo-RBBB and persistent ST segment elevation in leads V1 to V3. Brugada syndrome has 3 different patterns of ST elevation ECG pattern. The classic Brugada pattern (type 1) consists of ST-segment elevation ( $\geq 2$ mm) that descends with an upward convexity to an inverted T wave. This is referred to as the “coved type” Brugada pattern.

Type 2 and 3 patterns have a “saddle back” ST-T wave configuration, in which the elevated ST segment descends toward the baseline, then rises again to an upright or biphasic T wave. The ST segment is elevated  $\geq 1$  mm in type 2 and  $< 1$  mm in type 3.<sup>11</sup>

The differential diagnosis of Brugada syndrome includes anteroseptal myocardial infarction, hypercalcemia, hyperkalemia, left ventricular hypertrophy, pericarditis, and right bundle branch block.

In symptomatic patients, Implantation of Cardiac Defibrillator (ICD) is the only effective intervention for preventing sudden cardiac death. Asymptomatic individuals with spontaneous Brugada ECG are also at high risk for arrhythmic events if sustained ventricular arrhythmias are induced during Electrophysiological Study (EPS) and ICD is recommended, while asymptomatic patients have a benign prognosis if the EPS is negative and close follow up is recommended.<sup>12</sup>

In summary, Brugada syndrome is a recognized cause of sudden death in an apparently normal heart and its recognition might prevent a fatal outcome. To the best of our knowledge, this is the first reported case of this syndrome in a Jordanian family.

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## متلازمة بروغادا: حالة مرضية في الأردن

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### الملخص

متلازمة بروغادا هي حالة مرضية جديدة عُرفت لأول مرة عام 1992، وهي أحد أسباب الوفاة الفجائية وتمتاز بتغيرات في تخطيط القلب الكهربائي.

تم تقلص حالة مرضية لمريض ذكر يبلغ من العمر ( 21 ) عاماً تم تحويله الى مستشفى الجامعة الأردنية كحالة احتشاء عضلة القلب بناءً على تغيرات في تخطيط القلب الكهربائي، إذ كان هناك ارتفاع في منطقة س-ت شبيهه بتلك التغيرات التي تحصل في احتشاء عضلة القلب.

أجري للمريض فحص انزيمات القلب وقسطرة القلب والشرايين حيث كانت النتائج كلها طبيعية، وبعد مراجعة تخطيط القلب تبين أن هذه المتغيرات تطابق متلازمة بروغادا.

تم إجراء تخطيط القلب الكهربائي لعائلة المريض وتبين وجود التغيرات نفسها في كل من والده وأخيه.

الكلمات الدالة: متلازمة بروغادا، موت فجائي، احتشاء عضلة القلب.