СИНДРОМ БРУГАДА: КОГДА СЕРДЦЕ ПЕРЕСТАЕТ БИТЬСЯ

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Типичный пациент с синдромом Бругада – это молодой, не имеющий сопутствующих заболеваний мужчина, с нормальными результатами общеклинического обследования и физикального исследования сердечно-сосудистой системы. Описывается клинический случай успешной сердечно-легочной реанимации после остановки сердца у больного с диагностированным лишь после реанимации синдромом Бругада 2 типа.

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Ключевые слова: синдром Бругада, остановка сердца, сердечно-легочная реанимация.

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Introduction

During recent years, Brugada syndrome (BS) has become relatively new distinguished electrocardiographical (ECG) entity characterized by ST elevation in right precordial leads V1-V3 [1]. According to Jurcevic and colleagues, right bundle block, initially being described as integral characteristic of this syndrome, does not always have to be present [2]. The typical Brugada patient is young, otherwise healthy male with normal general medical and cardiovascular physical findings.

We call the heart of a patient with BS the "Brugada heart". Although not always, patients can have prodromes in a form of Brugada signs, such as: syncope, breathing difficulties, convulsions, ventricular arrhythmias, abnormal heart rhythm, or previous cardiac arrest (CA) [3]. Unrecognized "Brugada heart" with its increased potential for producing arrhythmias, most commonly leads to sudden death. Application of implantable cardioverter defibrillator (ICD) is treatment of choice for recognized "Brugada heart" and it is aimed at primary and secondary prevention of malignant ventricular rhythm disturbances (pulseless ventricular tachycardia – PVT or ventricular fibrillation – VF) [4].

We present a case of successful resuscitation following "Brugada heart" arrest, where diagnosis of type 2 Brugada syndrome was made after cardiopulmonary resuscitation (CPR).

Case report

Under first degree emergency call, Belgrade Emergency Medical Service crew was sent to see 33 years old patient who suddenly lost consciousness at home, and had breathing difficulties accompanied with wheezing and rasping. Upon arrival, 8 minutes after call was collected, the crew found patient on the floor in a state of CAt (unconscious, without breathing or pulse), while his mother was delivering basic life support (BLS) measures: sternal compression and artificial ventilation. Defibrillator monitor showed VF. In accordance to Advanced Life Support (ALS) protocol for shockable rhythm, DC shock was delivered twice, reestablishing normal heart rhythm, circulation (palpable carotid artery pulse) and breathing. As patient was in post-resuscitation comma (GCS=8), he was intubated with endotracheal tube through which he breathed spontaneously, while oxygen was delivered through nasal cannula at rate of 6 LPM. Intravenous lines were established on both hands and patient was started on cold normalized saline and Ringer's solution at 30 ml/kg dose. The patient was then transferred to Emergency Center where he regained consciousness two hours later, and was extubated after 3 hours. Brain CT scan was performed and was described as normal.

Patient's family history does not contain any data on sudden death or coronary disease. Until now, patient was healthy, did not take any therapy and denied alcohol and drug abuse.

Patient was finally transferred to the Institute for Cardiovascular Diseases. Upon admission, patient was afebrile, with normal breath sound and without any accompanying findings on auscultation. Heart rhythm was normal; sounds were clear and with no murmurs. Arterial tension (AT) was 120/70 mmHg.

The chest X-ray was normal. ECG findings: sinus rhythm, ventricular rate 60/min, PR interval 156/min, flattened T wave at D3, rR' at V1-V2 with ST elevation (saddle back pattern) measuring 1 mm, as well as positive T wave at V1-V2 (Figure 1).

Patient underwent thorough cardiological workup: echocardiogram (Echo) showed somewhat increased left ventricle diastolic dimension (5.8 cm), normal systolic dimension (3.8 cm) without segmental kinetic disturbances, and ejaction fraction (EF) of 61%. Right ventricle and pericardium showed no abnormalities. Selective coronarography (SC) showed normal findings. Laboratory and serum electrolytes results were normal.

In the light of repeated episode of syncope that occurred during hospital stay, the absence of heart condition, and ECG-confirmed diagnosis of type 2 Brugada syndrome (Table 1), it was decided that the patient suffered from "Brugada heart". Therefore, Medtronik Entrust ICD-VR=Single-chamber implantable cardioverter-defibrillator with ventricular endocardial electrode Medtronic Sprint Quattro 6944 has been implanted under local infiltrative anesthesia. Follow-up ECG showed that the pacing capture was normal (Figure 2) and that the pacemaker was optimally programmed.

The patient was discharged with advice for continuation of the following therapy: analgesics if needed, bisoprolol 2.5 mg (tbl. Concor®), and 100 mg Acetylsalicylic acid (tbl. Cardiopirin®).

Table 1

ST segment abnormalities in leads V1–V3 [4]

	Type 1	Type 2	Type 3
J-wave amplitude	Negative	Positive or biphasic	Positive
T-wave	≥2 mm	≥2 mm	≥2 mm
ST-T configuration	Coved type	Saddle back	Saddle back
ST segment (terminal portion)	Gradually descending	Elevated ≥1 mm	Elevated <1 mm

1 mm=0·1 mV, the terminal portion of the ST-segment refers to the latter half of the ST-segment.

Table 2

Diagnostic Criteria for BS [8]

Major criteria
1. Presence of the ECG marker of Brugada syndrome in patients with structurally normal heart 2. Appearance of the ECG marker of Brugada syndrome after administration of sodium channel blockers
Minor criteria
1. Family history of sudden cardiac death 2. Syncope of unknown origin 3. Documented episodes of ventricular tachycardia/ventricular fibrillation 4. Positive programmed electrocardiostimulation test on ventricular tachycardia/ventricular fibrillation 5. Genetic mutations of ion channels (to be defined)

ECG = electrocardiographic.

Discussion

Brugada syndrome should be suspected in successfully resuscitated younger male without any previous structural heart condition, resembling our patient [1]. In our patient, the absence of any previous structural heart condition was proved by supplementary diagnostic methods (Echo and SC). Coronarographic and ultrasonic findings were normal confirming that "Brugada heart" is an example of ion channel disorder (chanellopathy) caused by an alteration in the trans-membrane ion currents that together constitute the cardiac action potential [5].

In 34% of cases, the cause of syncope and sudden death of patients with "Brugada heart" are malignant rhythm disturbances, in which early defibrillation is the only treatment option according to ALS algorithm [6]. The success of CPR in our patient was contributed by his mother who applied BLS measures before the arrival of EMS crew.

Pathognomonic diagnostic confirmation of BS is based on typical ECG changes during rest (persisting or intermittent). According to special Arrhythmia Working Group of the European Society of Cardiology [4], three types of patterns have been described in BS. After successful CPR of our patient, ECG picture of type 2 Brugada syndrome is registered during rest (Figure 1). One study conducted in the United States [7] has shown that the prevalence of type 2 Brugada syndrome among adults is 0.14%.

Apart from ECG findings, the presence of at least one major and one minor Gussak criteria has high sensitivity for diagnosis of BS [8] (Table 2). Our patient had one major criterion: the presence of the ECG marker of BS in patients with structurally normal heart; and two minor criteria: syncope of unknown origin and documented episodes of PVT/VF. Due to lack of availability of these methods in Serbia, the analysis of genetic mutations for ion channels I and the induction of ECG changes using intravenous class IA antiarrhythmics (procainamide, aimaline and flecainide) were not performed. The experience of other authors [9] show that the above-mentioned antiarrhythmics used in accordance to the recommended protocol can unhide or intensify ECG changes, therefore having not only diagnostic, but therapeutic significance.

Because of high malignant potential of "Brugada heart", the accurate diagnosis is very important. The most significant differential diagnostic entity [10] is arrythmogenic right ventricular dysplasia (ARVD) that can be discovered either by histopathological finding of transmural fibro-adipose change on autopsy, or surgically (ventriculotomy with limited resection of arrhythmogenic area). The question that remains unanswered is whether BS represents spectral part of ARVD.

The findings that are available to the present day suggest that due to high risk of sudden death, the treatment of choice in both symptomatic and asymptomatic Brugada patients is application of ICD as superior method to pharmacological prevention using antiarrhythmics [9]. This is especially true for patients with frequent syncope, VF or those who were already resuscitated due to CA. The dilemma rises in case of typical ECG picture of BS without family history, as in our patient. The recommendations suggest the protection of this kind of "Brugada heart" with ICD implantation due to risk of sudden death. The mortality rate among Brugada patients [1] is 0% after ICD implantation, 26% in those treated with antiarrhythmics, and 31% in untreated patients.

Conclusion

When "Brugada heart" stops beating, the success of CPR is contributed by early BLS and early defibrillation. Unrecognized "Brugada heart" may be significant cause of

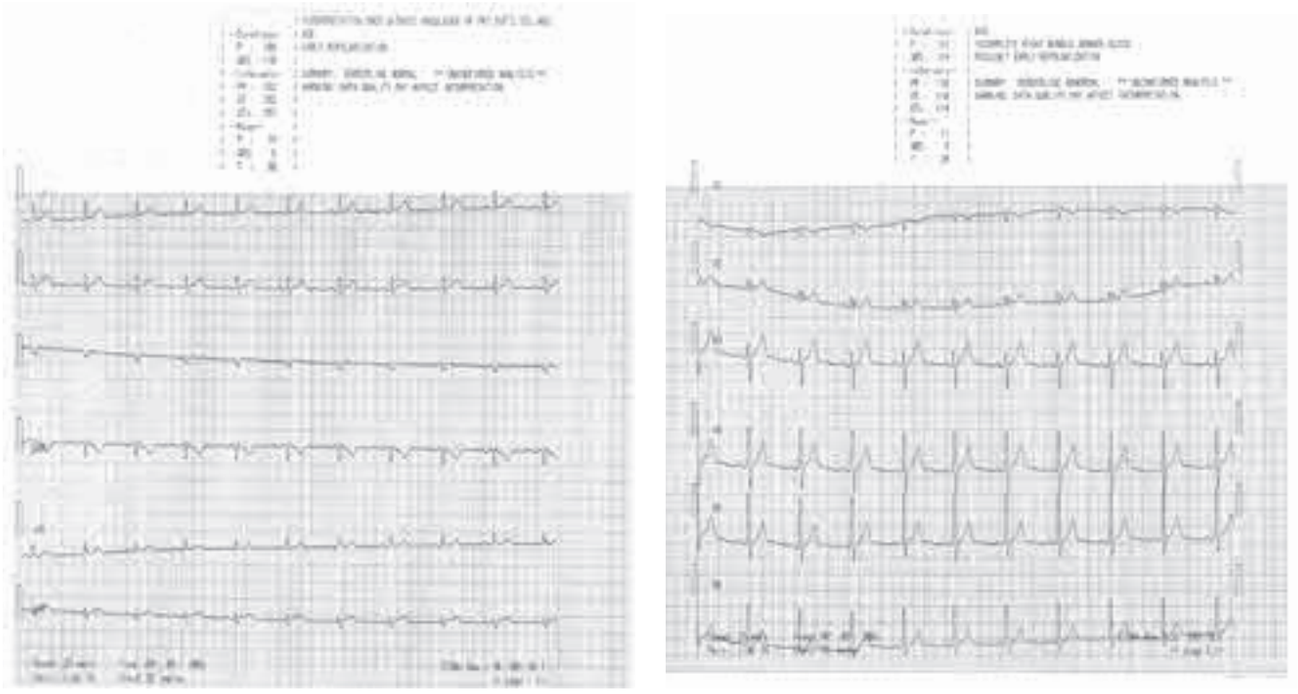

Figure 1. ECG picture of type 2 Brugada syndrome

Legend: ECG findings: sinus rhythm, ventricular rate 60/min, PR interval 156/min, flattened T wave at D3, rR' at V1-V2 with ST elevation (saddle back pattern) measuring 1 mm, as well as positive T wave at V1-V2.

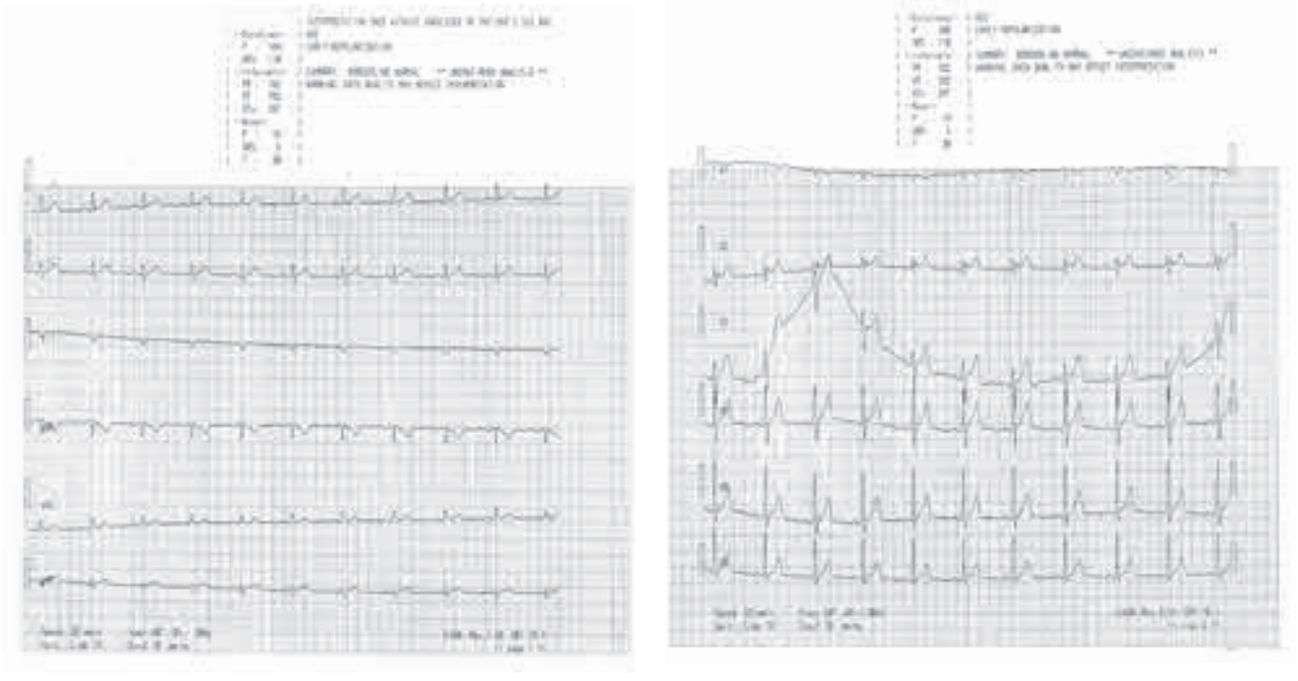

Figure 2. ECG finding after implantable cardioverter-defibrillator

Legend: ECG showed that the pacing capture was normal and that the pacemaker was optimally programmed.

death among males under 40. Diagnostic confirmation of "Brugada heart" is based on Brugada ECG pattern, absence of structural heart damage and the findings of ion channel disorder. Because of its recent identification, the prevalence of Brugada syndrome in Serbia is not well established. Due to high risk of sudden death, the treatment of choice in both symptomatic and asymptomatic Brugada patients is definitely ICD implantation.

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WHEN “BRUGADA HEART” STOPS BEATING

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The typical Brugada patient is young, otherwise healthy male with normal general medical and cardiovascular physical findings. A present a case of successful resuscitation following “Brugada heart” arrest, where diagnosis of type 2 Brugada syndrome was made after cardiopulmonary resuscitation (CPR).

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Key words: “Brugada heart”; cardiac arrest; cardiopulmonary resuscitation

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