Folia Cardiologica 2022
vol. 17, no. 2, pages 123–126
DOI: 10.5603/FC.2022.0017

ISSN 2353–7752
e-ISSN 2353–7760

Brugada syndrome: rare so, beware!

Zespół Brugadów — rzadkie schorzenie, więc zachowaj czujność!

Paweł Maciński, Andrzej Osiecki, Małgorzata Oleszczak-Kostyra, Wacław Kochman

Bielanski Hospital, Clinical Department of Cardiovascular Diseases, Centre of Postgraduate Medical Education, Warsaw, Poland

Abstract

Brugada syndrome (BrS) is a rare disease, caused by mutations in the gene that encodes cardiac sodium channel and is characterized by ST-segment elevation in the right precordial leads of electrocardiogram (ECG). This channelopathy significantly increases the risk of sudden cardiac death (SCD) due to life-threatening ventricular arrhythmias in individuals with structurally normal hearts. In this case report, the authors present a 60 years-old male after an episode of presyncope, with no personal or familial history of cardiovascular disease, whose spontaneous resting ECG showed Brugada pattern, which was confirmed in a provocative drug test with ajmaline.

Key words: Brugada syndrome, channellopathy, ventricular arrhythmias, sudden cardiac death, SCD, ajmaline

Folia Cardiologica 2022; 17, 2: 123–126

Introduction

In 1986, a young Polish boy with a recurrent syncopal attacks was referred to Brugada brothers — prominent cardiologists in Spain. The patient electrocardiogram (ECG) showed an ST-segment elevation in leads V_1 to V_3. His younger sister presented similar symptoms and ECG findings and died suddenly at the age of 2. In the following years of study, six additional cases came to their attention. In 1992 they described all cases as the groundwork for a novel clinical phenomenon [1]. Brugada syndrome (BrS) affects 1/2000 to 1/5000 individuals and occurs 8–10 times more frequently in men than in women [2]. Brugada syndrome is suggested to be involved in 4% to 12% of all sudden cardiac deaths (SCD) and at least 20% of sudden deaths in patients without structural heart disease. Although BrS may appear at any age, it is commonly found in young adults (20–39 years old) [3].

Case report

A white, 60 years-old male, ex-smoker (20 pack-years), with a history of radical prostatectomy (in 2018) due to prostatic adenocarcinoma, iatrogenic urinary incontinence, bilateral hearing loss, was admitted to the Department of Cardiology because of fatigue and progressive exertional dyspnea. The patient reported one episode of dizziness and presyncope while driving his car a few days before admission. He denied palpitations, chest pain, syncope, oedemas, orthopnea, and any other respiratory symptoms. There was no history of sudden death in any of his relatives. He reported, that over the last few years he had been referred several times to the E.R. with a suspicion of acute coronary syndrome due to ECG abnormalities, but the ischemic cause had always been ruled out. On admission the patient was in a good general condition, his physical examination, laboratory analysis, and chest X-ray were normal. The 12-lead ECG

Address for correspondence: Paweł Maciński MD, Klinika Chorób Układu Krążenia CMKP, Szpital Bielański Samodzielny Publiczny Zakład Opieki Zdrowotnej, ul. Cegłowska 80, 01–809 Warszawa, Poland, e-mail: pawel.macinski@icloud.com

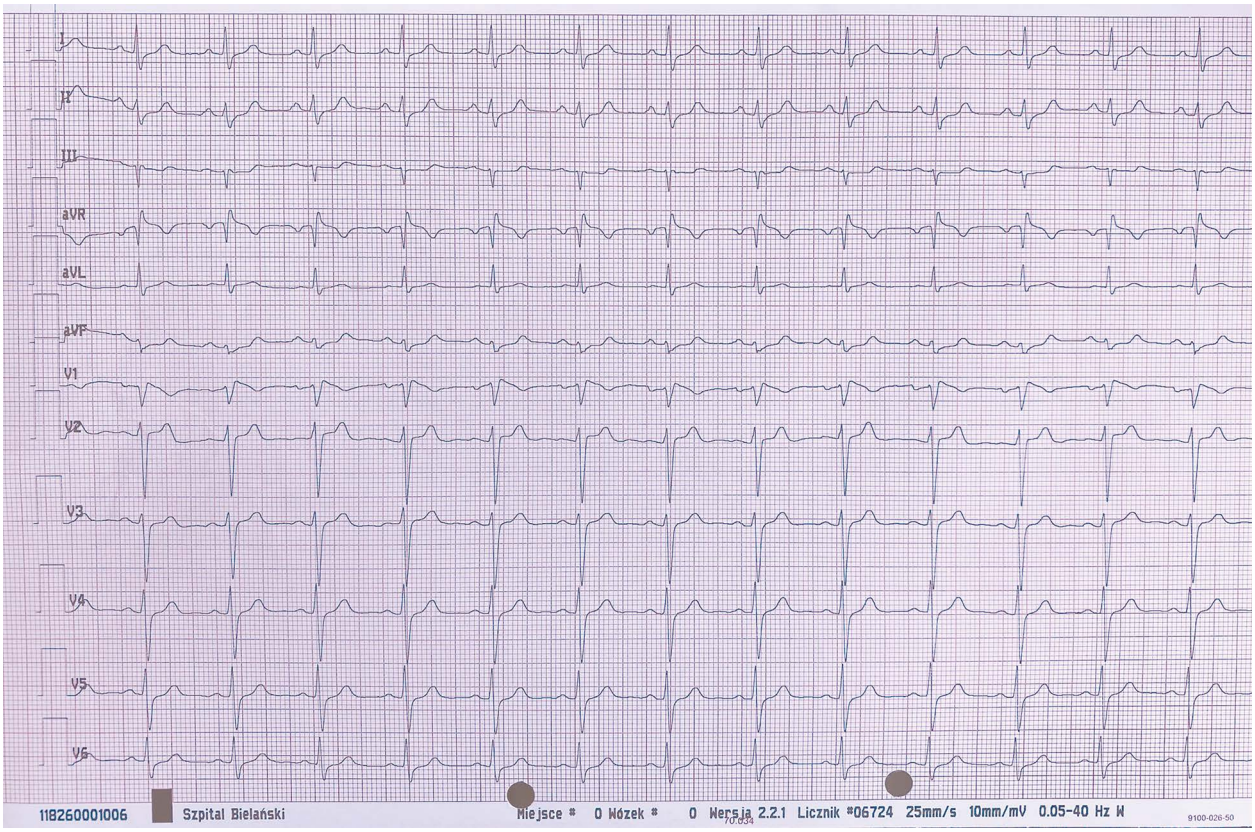

Figure 1. Spontaneous electrocardiographic changes suggesting Brugada type 1 pattern (coved-type ST-segment elevation in lead V_1)

on admission showed sinus rhythm 68 bpm, nonspecific intraventricular conduction delay (QRS width 120 ms), there was no evidence of cardiac ischemia. Transthoracic echocardiogram showed mild left ventricular hypertrophy, normal left ventricular systolic function (LVEF 56%), and no significant valvular defect. In 24-hours Holter monitoring the patient remained in sinus rhythm (average heart rate 73 bpm) with no significant supraventricular or ventricular arrhythmias. We decided to perform an exercise treadmill stress test using standard Bruce protocol as a diagnostic approach for coronary artery disease. The test was stopped at 3. minute of the protocol because of patients' chest pain and fatigue, at the workload level of 4.6 metabolic equivalents (METs), without concomitant ECG abnormalities. Subsequently, the patient underwent coronary angiography via radial approach, which showed normal coronary arteries. In order to extend diagnostic testing for exertion intolerance chest, CT scan was performed, which was normal, except for hepatic and renal cysts.

During the next days of hospitalization routine resting 12-leads ECG revealed changes typical for BrS (Figure 1).

A provocative drug test with ajmaline (intravenously in fractions 10 mg every 2 minutes, up to the target dose of 1 mg/kg body weight) was performed, and both ECGs (standard and obtained from the one intercostal space above) revealed ST-segment abnormalities characteristic for type 1 Brugada pattern with a 3 mm elevation in the right precordial leads (V_1 and V_2) (Figure 2). After 15 minutes ECG showed complete resolution of previous findings. During the drug test the patient was asymptomatic and hemodynamically stable.

In the light of a definitive diagnosis of Brugada syndrome type 1 based on a provocative drug test and the history of presyncope, the patient was qualified for cardioverter-defibrillator implantation (ICD) for primary prevention of SCD.

Discussion

Being an autosomal dominant channelopathy, BrS is mostly caused by a mutation in the gene SCN5A leading to a loss of function of the cardiac sodium channels, especially localized in the right ventricular outflow tract (RVOT). Patients may present with palpitations, difficulty in breathing, seizures, syncope, malignant arrhythmias (ventricular tachycardia/ventricular fibrillation) or SCD.

A diagnosis of BrS is established when ECG shows a "coved-type" ST-segment elevation (type 1) ≥ 2 mm in at least one right precordial lead (V_1 to V_2) in the II, III or IV intercostal space. These ECG pattern may occur spontaneously or after challenge with a sodium channel blocker (such as flecainide or ajmaline). Type 2 ("saddle-back type") is characterized by ST-segment elevation ≥ 0.5 mm

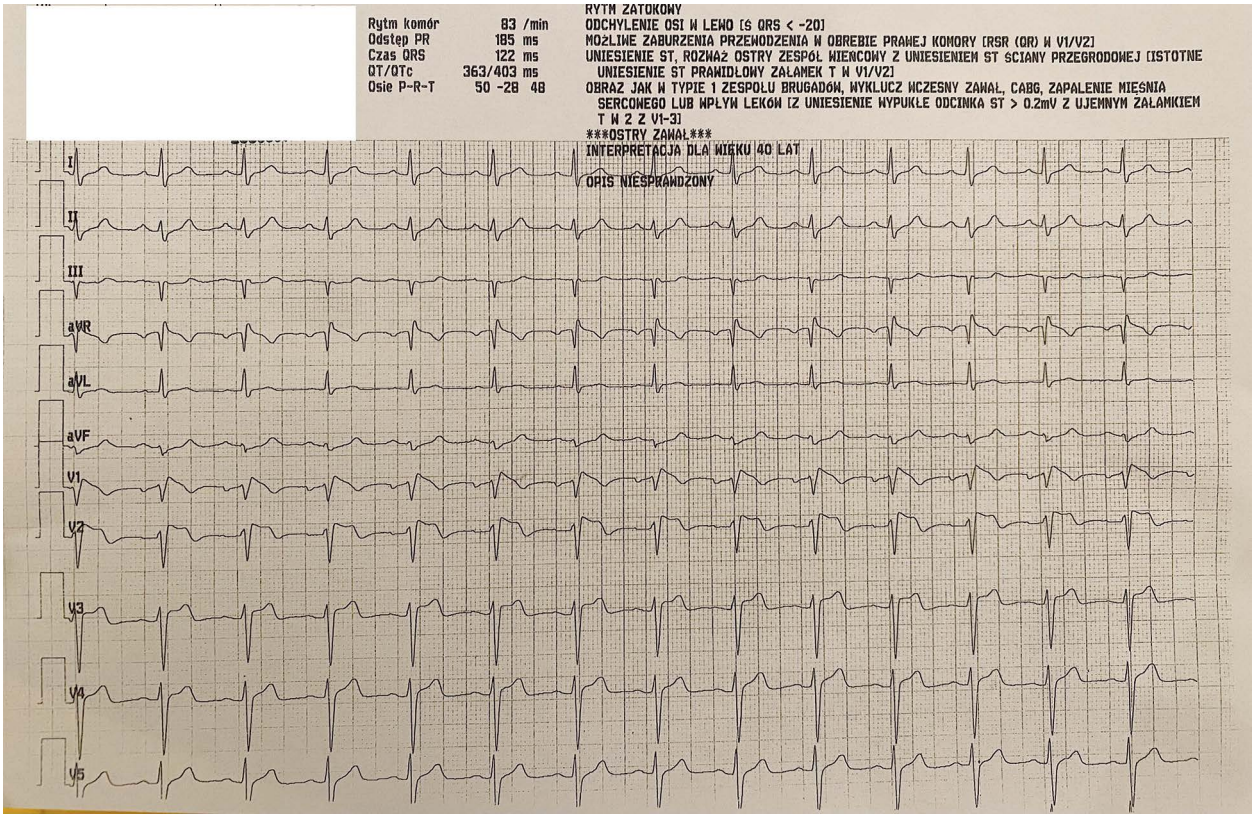

Figure 2. The standard electrocardiogram with positive ajmaline test result — ST-segment elevation 3 mm in leads V_1 and V_2

(generally ≥ 2 mm in V_2) in any of V_1 or V_2 lead, with a convex ST segment. Type 2 is non-diagnostic for BrS and should be differentiated from other Brugada-like ECG patterns occurring i.e. in athletes, patients with pectus excavatum, or arrhythmogenic right ventricular cardiomyopathy. Some additional criteria have been suggested to facilitate this differentiation. These criteria are based on the triangle formed by the ascending and descending branch of the r'-wave in the right precordial leads: β-angle ≥ 58° — the best predictor for conversion to type 1 BrS pattern (Figure 3A) and the triangle's base length ≥ 4 mm measured 5 mm below the peak of the r'-wave (Figure 3B) [4–6].

Malignant arrhythmias most commonly occur at rest or during sleep, which is related with physiological states of increased vagal tone. Furthermore, some substances and conditions (trigger factors) such as certain drugs, fever, excessive alcohol intake, and large meals can reveal or exacerbate ECG patterns [7]. All patients with BrS should be educated to avoid these factors.

An ICD is currently recommended for primary and secondary prevention of sudden cardiac death in patients with BrS. According to 2015 European Society of Cardiology guidelines for the prevention of SCD, an ICD is indicated in symptomatic patients (survivors of an aborted cardiac arrest and/or with documented spontaneous sustained ventricular tachycardia, also patients with a history of unexplained loss of consciousness, seizures, or agonal respiration at night). In asymptomatic patients with abnormal ECG findings, the electrophysiological study (EPS) may unmask Brugada-like pattern and we should also treat them with ICD [8]. Recently, epicardial catheter ablation has been reported as a novel therapeutic tool [8, 9]. However, longer follow-up is required before entering it into the general clinical practice.

Conclusions

It should be borne in mind that Brugada electrocardiographic pattern is dynamic and can be concealed. In this case, we highlight the need for physicians to be very watchful in identifying patients with Brugada syndrome to prevent malignant arrhythmias and lower the risk of SCD.

Conflict of interest

The authors declare that there is no conflict of interest.

Funding

No funding sources.

Ethics

Ethical approval is not required at the authors' institution to publish an anonymous case report.

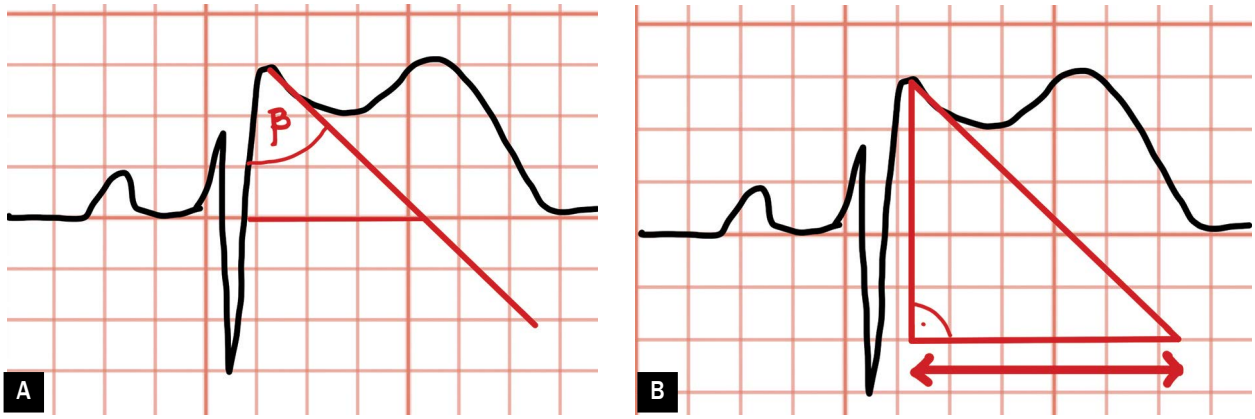

Figure 3A. The β-angle in right precordial lead; appears between ascending and descending branch of the r'; **B.** The length of the triangle base measured 5 mm below the peak of the r'-wave in the right precordial lead

Streszczenie

Zespół Brugadów jest rzadkim schorzeniem, spowodowanym mutacjami genu kodującego sercowe kanały sodowe, charakteryzującym się uniesieniem odcinka ST w prawokomorowych odprowadzeniach przedsercowych w zapisie elektrokardiograficznym (EKG). Kanałopatia ta wiąże się z istotnie zwiększonym ryzykiem wystąpienia nagłej śmierci sercowej w przebiegu złośliwych arytmii komorowych u pacjentów bez strukturalnej choroby serca. W niniejszym artykule autorzy przedstawiają przypadek kliniczny 60-letniego mężczyzny po przebytym stanie przedomdleniowym, bez rozpoznanej choroby sercowo-naczyniowej i z nieobciążającym wywiadem rodzinnym, u którego w EKG zarejestrowano zmiany sugerujące zespół Brugadów, co zostało potwierdzone w teście prowokacji farmakologicznej ajmaliną.

Słowa kluczowe: zespół Brugadów, kanałopatia, arytmie komorowe, nagła śmierć sercowa, ajmalina

Folia Cardiologica 2022; 17, 2: 123–126

References

1. Brugada P, Brugada J. Right bundle branch block, persistent ST segment elevation and sudden cardiac death: a distinct clinical and electrocardiographic syndrome. J Am Coll Cardiol. 1992; 20(6): 1391–1396, doi: 10.1016/0735-1097(92)90253-j, indexed in Pubmed: 1309182.
2. Quan XQ, Li S, Liu R, et al. A meta-analytic review of prevalence for Brugada ECG patterns and the risk for death. Medicine (Baltimore). 2016; 95(50): e5643, doi: 10.1097/MD.0000000000005643, indexed in Pubmed: 27977610.
3. Antzelevitch C, Brugada P, Borggrefe M, et al. Brugada syndrome: report of the second consensus conference: endorsed by the Heart Rhythm Society and the European Heart Rhythm Association. Circulation. 2005; 111(5): 659–670, doi: 10.1161/01.CIR.0000152479.54298.51, indexed in Pubmed: 15655131.
4. Brugada J, Campuzano O, Arbelo E, et al. Present status of Brugada syndrome: JACC State-of-the-Art review. J Am Coll Cardiol. 2018; 72(9): 1046–1059, doi: 10.1016/j.jacc.2018.06.037, indexed in Pubmed: 30139433.
5. Chevallier S, Forclaz A, Tenkorang J, et al. New electrocardiographic criteria for discriminating between Brugada types 2 and 3 patterns and incomplete right bundle branch block. J Am Coll Cardiol. 2011; 58(22): 2290–2298, doi: 10.1016/j.jacc.2011.08.039, indexed in Pubmed: 22093505.
6. Serra G, Baranchuk A, Bayés-De-Luna A, et al. New electrocardiographic criteria to differentiate the type-2 Brugada pattern from electrocardiogram of healthy athletes with r'-wave in leads V1/V2. Europace. 2014; 16(11): 1639–1645, doi: 10.1093/europace/euu025, indexed in Pubmed: 24603955.
7. Tomé G, Freitas J. Induced Brugada syndrome: possible sources of arrhythmogenesis. Rev Port Cardiol. 2017; 36(12): 945–956, doi: 10.1016/j.repc.2017.06.015, indexed in Pubmed: 29233646.
8. Priori SG, Blomström-Lundqvist C, Mazzanti A, et al. ESC Scientific Document Group. 2015 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death: The Task Force for the Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death of the European Society of Cardiology (ESC). Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC). Eur Heart J. 2015; 36(41): 2793–2867, doi: 10.1093/eurheartj/ehv316, indexed in Pubmed: 26320108.
9. Pappone C, Brugada J, Vicedomini G, et al. Electrical substrate elimination in 135 consecutive patients with Brugada syndrome. Circ Arrhythm Electrophysiol. 2017; 10(5): e005053, doi: 10.1161/CIRCEP.117.005053, indexed in Pubmed: 28500178.