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TRANSIENT BRUGADA SYNDROME IN CLINICAL PRACTICE OF UKRAINE

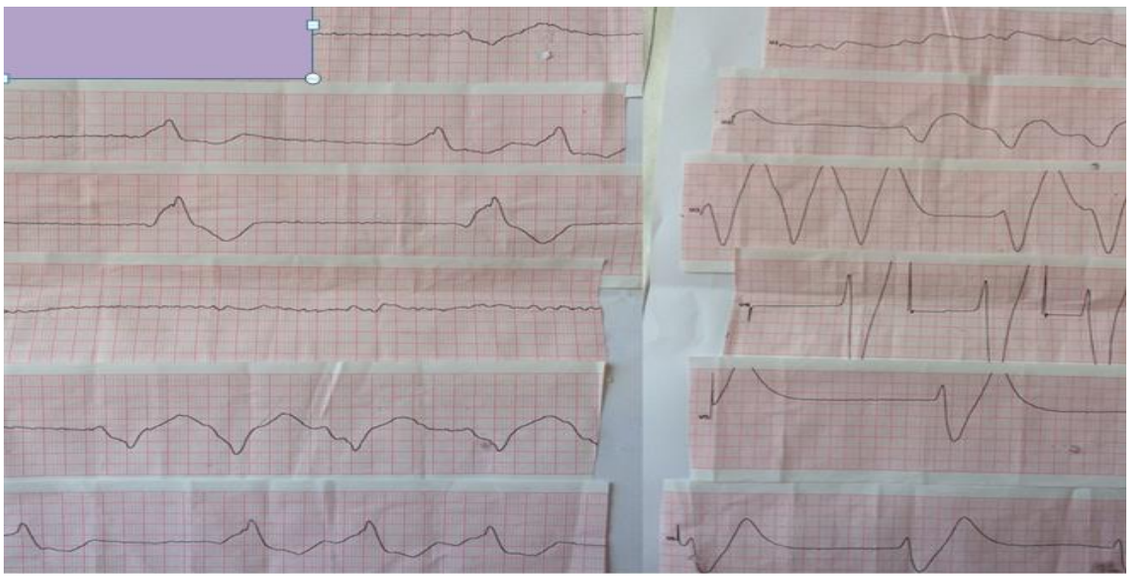
Abstract. *The article presents the clinical case of the Brugada syndrome, which had a transient course, to consider modern approaches to diagnosis and tactics of its objectivization.*
Key words: *Brugada syndrome, diagnostics.*

Introduction. In 1992, the P. and J. Brugada brothers described a new clinical and electrocardiographic (ECG) syndrome for the first time, characterized by Right bundle branch block, persistent Elevation of the ST segment in the right chest leads, and Sudden cardiac death (CRC) [4]. The authors presented data on 8 patients (6 men and 2 women) with repeated episodes of transient sudden death, the main cause of which was polymorphic ventricular tachycardia (VT) with a large frequency of ventricular contractions provoked by ventricular extrasystole. Brugada syndrome (BRS) is considered as one of the main causes of CRC in young patients without cardial pathology. The important fact is that the literature describes significant ethnic and geographic variations in the population of patients with BRS. The prevalence of BRS with ECG manifestations [3] is higher in adults in Asian countries such as Japan (0.15-0.27%), Philippines (0.18%) than in Western countries, including Europe (from 0 to 0.017%) or North America (0.005-0.1%) [5].

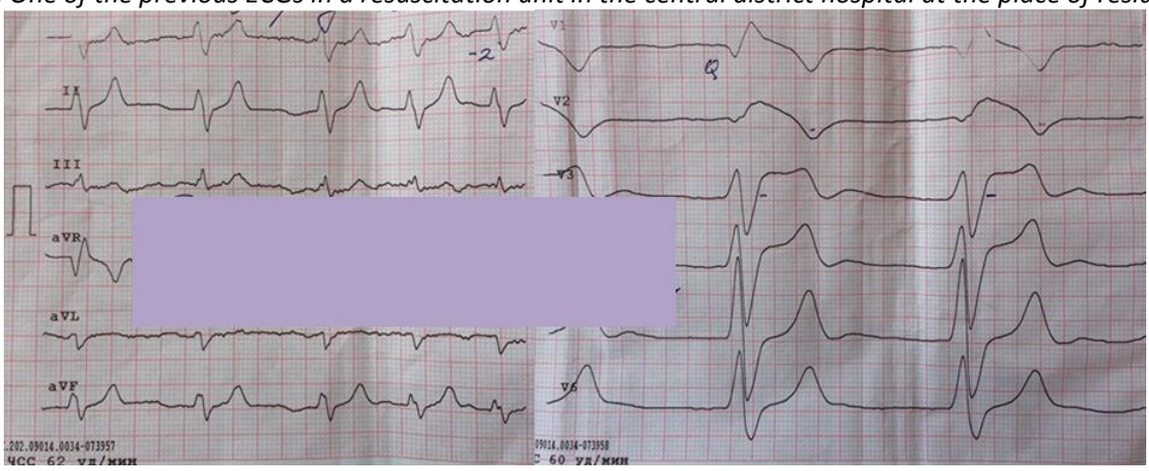
Transient BRS (type 1) can be triggered by the use of antiarrhythmic drugs (AAP) from the group of blockers of sodium channels, psychotropic drugs, fever, electrolyte disorders – high potassium (hyperkalaemia), alcohol or intoxication with cocaine, etc. Other reasons for the elevation of the ST segment, which should be excluded before the diagnosis of BRS, is the complete of the right bundle branch block, arrhythmogenic cardiomyopathy of the right ventricle, occlusion of the left anterior coronary artery, etc. [5].

Material and methods. Here is a clinical case of diagnosis of transient BRS. Patient G. born in 1944 (74 years old), was admitted to the in-patient treatment on 05.06.2018 in the Regional Clinical Cardiologycal Center in Chernivtsi with complaints about periodic compression of the pain behind the sternum, which was not caused by physical activity, general weakness, dizziness, feeling of interruptions in the work of the heart, and periodic feeling of "lack of air". It is known from the anamnesis that she was treated a month ago in the intensive care unit of the central district hospital for instability of arterial pressure, paroxysmal tachyarrhythmia and episodes of loss of consciousness (2 cases during the last month). At the previous archival ECG, one of which is presented in picture 1, atrial fibrillation, episodes of accelerated idioventricular rhythm, short ventricular tachycardia, and even an episode of ventricular fibrillation which was spontaneously renewed. Diagnostic for BRS is family history with syncope, genetic test for mutation of BRS, etc. [6].

After the treatment in the central hospital, the patient's condition improved slightly, for further treatment and diagnostic was directed to the Regional Clinical Cardiologycal Center in Chernivtsi, where he was urgently hospitalized in the department of acute coronary insufficiency to stabilize the condition. Objective examination: general condition of the patient of moderate severity, Pulse - 60 beats per min, arrhythmic, BP - 140/80 mm Hg. There is vesicular breathing and no wheezing in the lungs. On the ECG (picture 2), the direction of the ST segment in the shape of a characteristic "vaulted" oblique-basement elevation in the right thorax V1-V3 to 3-5 mm, that passed into the negative symmetrical T-wave in these leads, QT extended in the right chest leads, and therefore certain changes required further differentiation [1], also the electric axis of the



Pic. 1. One of the previous ECGs in a resuscitation unit in the central district hospital at the place of residence.



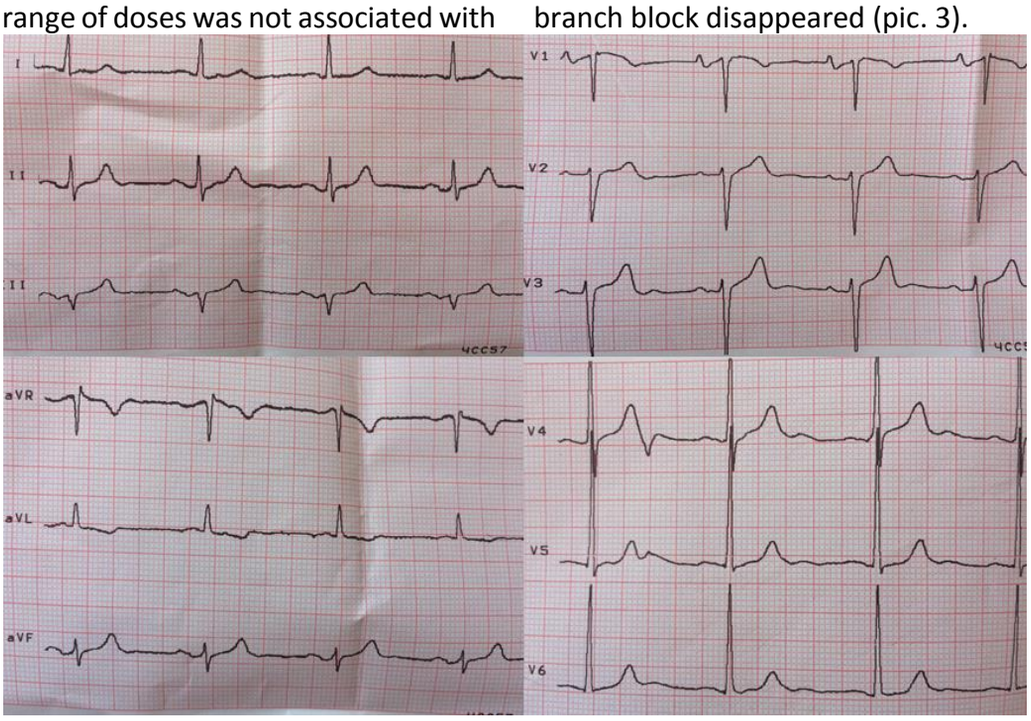
Pic. 2. ECG when the patient was admitted to the Regional Clinical Cardiology Center.

heart was rejected to the right, Right bundle branch block, the transient arrhythmic component of the ECG was recorded - episodes of premature supraventricular beats and short areas of supraventricular tachyarrhythmia, atrial fibrillation. Echocardiography showed that the final diastolic size of the left ventricle (LV) was 5.4 cm, the ultimate systolic LV size was 3.8 cm, the ejection fraction (EF) was 56%, the left atrium (LA) was 4.8 cm, the thickness of the interventricular septum in diastole was 1.2 cm, the thickness of the posterior wall of the LV in diastole was 1.3 cm, the right ventricle was the standard size, the diameter of the aorta is 2.9-3.3 cm, the opening of the aorta valves was 1.0 cm , the aortic valve (AV) is considerably grafting, the regurgitation flow to AV I-II, the regurgitation stream was registered with the mitral valve (MV) I-III and at the tricuspid valve (TV) III. Consequently, at the time of arrival, the acute form of sites of hypokinesis was not found out, the conclusion of echocardiography - aortic atherosclerosis, degenerative sensation of the faces of the AV, stenosis of the AV, insufficiency of AV II and MV II-III st and TV III, hypertrophy of the LV, increase of the cavity of LA, EF was slightly lowered within the age limit. Blood analysis for troponin T was negative, repeated after 12 hours was also negative. Total blood count: erythrocytes - $3{,}58 \times 10^{12}$/L, hemoglobin 110 g/L, color index 0,92, platelets 212×10^{9}/L, leukocytes $5{,}9 \times 10^{9}$ /L. In the biochemical analysis of blood: glucose 5.9 mmol/L, creatinine 116.6 μmol/L, total cholesterol 3.6 mmol/L, total bilirubin 13.6 mmol/L, potassium 5.5 mmol/L, alanine aminotransferase 0,65 U/L, aspartate aminotransferase 0,31 U/L, fibrinogen 2.97 g/L, international normalized ratio 1.10, prothrombin index 92%.

Research results. Anti-ischemic treatment was prescribed, including the use of prolonged nitrates, antithrombotic agents, statins, and AAPs (in the short-term, the combination of AAPs II and III classes - bisoprolol (because randomized controlled trials (RCTs) had not been confirmed, evidence was

contradictory) and amiodarone (there is no risk in RCTs or very little evidence according to www.brugadadrugs.org/pref_avoid/, although classified as "drugs better avoided") etc.

According to [7] modern approaches to beta-blockers in BRS, their long-term treatment within the normal range of doses was not associated with exacerbation of clinical parameters and ECG results. In our own study, the patient condition had been improved, there was a normalization of ECG-indicators - sinus rhythm was renewed, tendency to sinus bradycardia with heart rate was recorded 55-60 beats per min, Right bundle branch block disappeared (pic. 3).



Pic. 3. ECG after received treatment.

It should be noted, that the best AAP is currently available in the clinic for BRS remains quinidine [5], provocative continue to be considered flekainid, procainamide, aimalin [6], class AAP IC, calcium channel blockers and amiodarone also show contraindications, beta-blockers are associated with the increased risk of developing ventricular arrhythmias in some patients with BRS, the proportion of high-risk patients with BRS requires implantation of cardioverter defibrillators, and there is a risk of their response to accelerated arrhythmias, and thus according to the authors [8], effect can be achieved by the introduction of quinidine at BRS without mutations in the SCN5A, including taking into account its effects in the study QUIDAM.

Thus, BRS, was first registered in 1992, is an inherited arrhythmic disease associated with ventricular fibrillation, premature sudden arrhythmic death, which has an autosomal dominant inheritance, which involves more than 100 mutations and 30% mutations exclusively of the SCN5A gene [9], which encodes the protein structure of the alpha subunit of sodium channels, which provides the sodium current of the potential of action. The syndrome is characterized by inactivation of sodium channels with the described specific characteristics of changes in the segment ST (signs of Right bundle branch block) with the rise of segment ST ("J Wave"-syndrome) in leads V1-V3 without evidence of ischemia, electrolyte imbalance and other structural heart diseases [16]. It is recognized that BRS is an inherited sodium channel pathology, which corresponds to 4-12% of sudden coronary death cases in young men who have tendentious to ventricular arrhythmias [9] and at least 20% of deaths in patients with structurally normal hearts [11].

In the Ukrainian recommendations [2] BRS is considered as 1) clinical criteria (syncope, sudden death), 2) ECG criteria (Right bundle branch block, ST elevation greater than 0.1 mm in leads V1-V3, ventricular arrhythmias- ventricular tachycardia and ventricular fibrillation), 3) absence of other pathology of the heart that could cause these clinic-electrocardiographic changes. Thus, according to the submitted personal data, an older age group female patient, with a potassium content that does not meet the criterion of hypercalemia (6.0-8.8 mmol/L) for BRS [6] was examined, in contrast to the presented conditions

in the differentiation of ECG changes with acute myocardial infarction [12], and the J-wave has recently been associated with inherited vital arrhythmic disorders, which include BRS and early repolarization syndrome as a group of "J Wave"-syndromes [5, 13].

A more differentiated approach was developed according to the consensus of the "J Wave"-syndrome, when the Shanghai Brugada Scoring System (SBSS) was proposed by the HRS / EHRA / APHRS / SOLAECE in 2016. The system takes into account ECG registration, genetic outcomes, clinical characteristics, and family history [14] with the definition of "probable" and/or "definite" BRS, "possible" BRS and "non-diagnostic" result in the distribution ≥3.5, 2-3 and <2 points, respectively.

The SBSS index predominated 6.5 in our patient (without genetic testing), and thus testified to a "certain and/or probable" BRS and a high risk of arrhythmic event, and the need for an ICDs because BRS (type 1) or atypical elevation of the ST segment in the right chest leads is caused by an increased risk sudden coronary death of middle-aged patients [15].

Conclusion. The most important is the objectification of BRS for differentiating the electrocardiograms of STEMI vs "J Wave" syndromes and determining the effective AAP-correction.

References.

1. Pavlenko TA, Blagova OV. Brugada syndrome: from primary electrical heart disease to the morphological substrate. Archive of Internal Medicine.2016; 2:61-69.

2. Sichov OS, Lutai MI, Romanova OM. Ambulatory Holter monitoring of ECG (draft recommendations). Ukrainc'kyi kardiolohichnyi zhurnal. 2005; 5:11-36.

3. Tashchuk VK, Polyanska OS, Ivanchuk PP. etc. Electrocardiography: clinical features and achievements in the historical aspect. Clinical anatomy and operative surgery.2015;14(3):123-134.

4. Brugada P, Brugada J. Right bundle branch block, persistent ST segment elevation and sudden cardiac death: a distinct clinical and electrocardiographic syndrome. A multicenter report. JACC, 1992; 15; 20(6):1391-1396.

5. Di Diego J.M., Antzelevitch C. J Wave Syndromes as a cause of Malignant Cardiac Arrhythmias. Pacing Clin. Electrophysiol. 2018 Jun 5. doi: 10.1111/pace.13408. [Epub ahead of print].

6. Dendramis G., Petrina S.M., Baranchuk A. Not all ST-segment elevations are myocardial infarction: Hyperkalemia and Brugada phenocopy. Am J Emerg Med. 2017; 35(4):662.

7. Kamakura T, Wada M, Ishibashi K. Feasibility evaluation of long-term use of beta-blockers and calcium antagonists in patients with Brugada syndrome. Europace.2018;20(I1):f72-f76.

8. Halperin L, Mellor G, Talajic M. Quinidine effective for the management of ventricular and atrial arrhythmias associated with Brugada syndrome. Heart Rhythm Case Rep. 2018;4(7):270-2.

9. Baum B, Ingaramo O, Chao D. Adolescent Seizure in the Emergency Department Due to Concomitant Brugada Syndrome. Pediatr Emerg Care. 2018;34(8):1-3.

10. Vutthikraivit W, Rattanawong P, Putthapiban P. Worldwide Prevalence of Brugada Syndrome: A Systematic Review and Meta-Analysis. Acta Cardiol. Sin.2018;34(3):267-77.

11. Asvestas D, Tse G, Baranchuk A. High risk electrocardiographic markers in Brugada syndrome. Int J Cardiol Heart Vasc.2018;18:58-64.

12. De Bliek EC. ST elevation: Differential diagnosis and caveats. A comprehensive review to help distinguish ST elevation myocardial infarction from nonischemic etiologies of ST elevation // Turk J Emerg. Med. 2018;18(1): 1-10.

13. Priori SG, Napolitano C. J-Wave Syndromes: Electrocardiographic and Clinical Aspects. Card Electrophysiol Clin. 2018;10(2):355-69.

14. Kawada S., Morita H., Antzelevitch C. et al. Shanghai Score System for Diagnosis of Brugada Syndrome: Validation of the Score System and System and Reclassification of the Patients. JACC Clin. Electrophysiol. 2018; 4(6):724-30.

15. Tsuneoka H, Takagi M, Murakoshi N. Long-Term Prognosis of Brugada-Type ECG and ECG With Atypical ST-Segment Elevation in the Right Precordial Leads Over 20 Years: Results From the Circulatory Risk in Communities Study (CIRCS). J Am Heart Assoc. 2016; 5(8):1-9.

16. Vutthikraivit W., Rattanawong P., Putthapiban P. Worldwide Prevalence of Brugada Syndrome: A Systematic Review and Meta-Analysis. Acta Cardiol Sin. 2018;34(3):267-77.