

BRUGADA SYNDROME — A CASE REPORT

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Abstract: Brugada syndrome is a type of arrhythmia disorder, which is characterised by abnormal electrocardiogram (ECG) findings and an increased risk of sudden cardiac death. The most frequent sign is a persistent ST elevation in the electrocardiographic leads V₁-V₃ with a right bundle branch block (RBBB). We present a case of 12 years old healthy child, without any complains until then. He had 2 episodes of collapse/syncope, which lasted long and spontaneously disappeared. The collapses were provoked by physical activity. On ECG we found sinus rhythm 62 bpm, RBBB (right bundle brunch block) and Brugada signs in V₂ and V₃ channel-ST elevation ≥ 2 mm. The child was sent in electrophysiological centre abroad where the electrophysiological study was performed. They did not found any accessory pathway. The atrioventricular (AV) conduction was normal. Long lasting polymorphic ventricular tachycardia/fibrillation was induced with prograded stimulation with 3 extrastimuli in right ventricular outflow tract. Performing one defibrillation the rhythm turned in sinus way. Then they performed ECG with translocation of electrodes V₁₋₃ in 2nd intercostal space and the Brugada I type findings was discovered. After confirming of presence of Brugada type -1 syndrome the implantable cardioverter-defibrillator (ICD) was applied on child heart.

Key words: Brugada syndrome, ventricular fibrillation, sudden cardiac death.

INTRODUCTION

Brugada syndrome is a type of arrhythmia disorder, which is characterised by abnormal electrocardiogram (ECG) findings and an increased risk of sudden cardiac death. It is the major cause of sudden death in adults and most common reason of unexplained death in young men without known underlying cardiac disease (1).

First time was reported in 1989, but in 1992 was for the first time recognized and described as a new clinical entity and named Brugada (by the brothers Paedro and Joseph- Spanish cardiologists). The syndrome is mostly genetic disorder, frequently causes cardiac death due to ventricular fibrillation. A persistent ST elevation (≥ 2 mm) in the electrocardiographic leads V₁-V₃ with a right bundle branch block (RBBB), with or without the terminal S waves in the lateral leads that are associated with a typical RBBB is the most frequent sign of recognitions the Brugada syndrome. This abnormality — called a type 1 Brugada ECG pattern — is detected only by an electrocardiogram (2, 3).

Brugada syndrome is much more common in men. The typical patient with Brugada syndrome is young (under 40 years) male, and otherwise healthy person, with normal general medical and cardiovascular physical examinations. Many people who have Brugada syndrome don't have any symptoms, and so they're unaware of their condition (4).

Brugada-type ECG isn't frequent. It has been identified in the United States in 0.012% and in Canada in 0.07% (5). The prevalence of type 1 ECG in the healthy Asian population is considered to be around 0.15% in adults and 0.005% in children. However, it is considered to be less than 0.02% in the Western population (6, 7).

There are not many reports in the literature data about this syndrome in children.

CASE REPORT

We present a case of 12 years old healthy child, without any complains until then. He had two episodes of collapses/syncope, which lasted long and spontaneously disappeared. The collapses were provoked by physical activity.

Familiar anamnesis was negative; none of the relatives had any medical history of cardiac disease. No data of sudden cardiac death in the family, two generations back.

On physical examination we had a child in very good condition, with normal physical growth and development (weight 42 Kg). Heart rhythm was rhythmical with 60/ beats per minute, normal heart sounds (first and second heart tone), no evidence of murmur, good pulses on 4 extremities, without organ enlargement. The blood pressure was normal 120/70 mm Hg.

The chest X-ray showed normal heart silhouette and normal lung vascularisation.

On ECG we found sinus rhythm 62 bpm, RBBB (right bundle brunch block) and Brugada signs in V_1 and V_2 channel- ST segment elevation ≥ 2 mm (electrodes positioned in 2nd, 3rd and 4th intercostal space) (Figure 1).

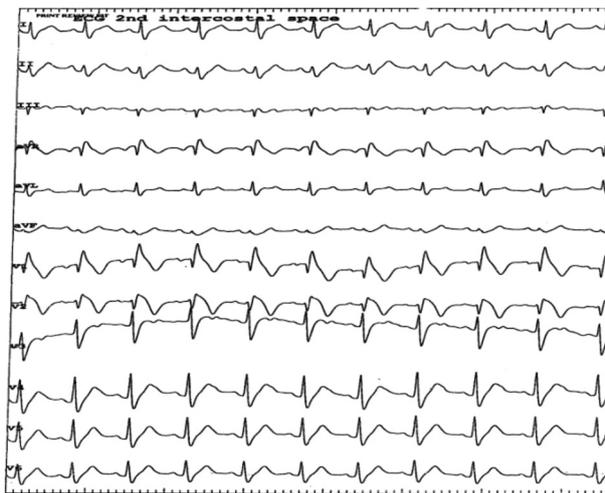


Figure 1. ECG in a child with Brugada syndrome

Echocardiogram was normal (normal structure; ejection fraction (EF) = 69% and Fraction shortening (FS) = 39%; normal morphology and function of the AV and semilunar valves, no evidence of septal defects).

Holter 24 hour ECG showed Brugada type findings, but without presence of ventricular tachycardia or fibrillation. On laboratory tests we found normal potassium and calcium levels in the serum, also CK-MB fraction in serum was normal.

The child was sent in electrophysiological centre abroad where the electrophysiological study was performed. They did not found any accessory pathway. The AV conduction was normal. Long lasting polymorphic ventricular tachycardia/fibrillation was induced with programmed stimulation with 3 extras stimuli in right ventricular outflow tract. By performing one defibrillation the rhythm turned in sinus way. Then they performed ECG with translocation of electrodes

V_{1-3} in 2nd intercostal space and the Brugada 1. type findings were discovered.

After confirming of presence of Brugada type -1 syndrome the implantable cardioverter- defibrillator was applied on child chest and connected by electrodes on the heart (Figure 2).

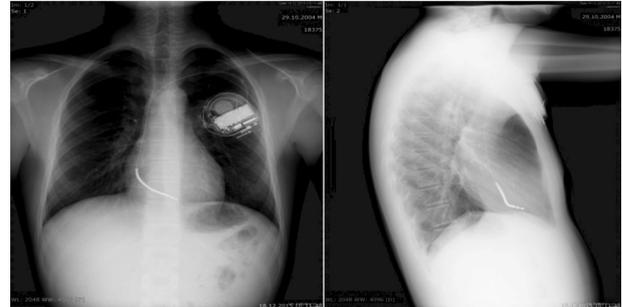


Figure 2. Xray of the chest – visualisation of intra-cardiac converter/defibrillator

The child had regularly check-up (according the ESC 2015 guidelines) with ECG, Holter 24 hour ECG and echocardiography 1 month after application of ICD, and every 3 month after that. After three years follow up the child was in very good condition, with normal physical activity, no evidence of infection, three short episodes of tachycardia at home (probable ventricular tachycardia/fibrillation) were stopped with the shocks from the device. On ECG and Holter ECG we did not found ventricular tachycardia/fibrillation. The echocardiography was normal. He is under therapy with Metoprolol retard (Beta blockers therapy).

This is the unique patient-child in our country with confirmed diagnosis of Brugada syndrome, and first and unique child with applied ICD.

DISCUSSION

Three different types of ECG patterns in Brugada syndrome are known. A type 1 ECG pattern is characterised by pronounced elevation of the J point (arrow), a coved-type ST segment, and an inverted T wave in V_1 and V_2 . A type 2 pattern is with a saddleback ST-segment elevated by >1 mm (usually is seen in healthy subject). A type 3 pattern has either a coved or a saddle back pattern with less than 2 mm J-point elevation and less than 1 mm ST segment elevation (8). Signs and symptoms in patients with Brugada syndrome may include the following:

- Syncope and/or cardiac arrest: Most common clinical manifestations; in many cases, cardiac arrest occurs during sleep at night or rest
- Nightmares or thrashing at night
- Asymptomatic, but routine ECG shows ST-segment elevation in leads V_1 - V_3

- Associated atrial fibrillation (20%)
- Fever: frequently reported to be a trigger or exacerbate clinical manifestations

The lack of a prodrome has been reported to be more common in patients with ventricular fibrillation documented as the cause of syncope in patients with Brugada syndrome (9).

Literature data reports that sudden cardiac death or ventricular fibrillation occurred in 8.2% of patients with Brugada syndrome. A history of syncope, a spontaneously abnormal ECG, and inducibility during programmed electrical stimulation (by one study) significantly increased this risk (10).

All patients with suspected Brugada syndrome need the following: 12- leads standard ECG and electrophysiological study for determination the inducibility of life threatening arrhythmias for risk stratification. In some cases may be performed laboratory test, which may be helpful for the diagnosis such as potassium and calcium levels, CK-MB and troponin levels, and genetic testing for a mutation in SCN5A gene (11).

It is necessary to perform imaging studies - echocardiography and/or magnet resonance imaging primary to excluding other reasons for potential life threatening arrhythmias like arrhythmogenic right ventricular dysplasia, Duchenne muscular dystrophy, acute myocarditis, infarction of the myocardium (12).

Because of genetic origin of the disorder (autosomal dominant inheritance), patient with Brugada syndrome may be genetically tested for a mutation of SCN5A. This mutation affects the cardiac sodium channel subunits or proteins that regulate them. Literature data showed that only in 11-28 % of the population the genetic test of mentioned SCN5A gene is proven (negative genetic test did not exclude the disorder) (13).

Brugada syndrome is 8-10 time more prevalent in men than in women, but there is not difference of carrying mutation in both sex. The penetrance of the mutation is probable higher in man than in women. Brugada syndrome may affect individuals of any age (0-83), but most often the symptoms occur around the age of 40. Brugada syndrome is known as a sudden unexpected nocturnal death syndrome (SUNDS). Also is presented as a sudden infant death syndrome (SIDS), which means death in infant (within first year of life) without any previous cause or disorder (11, 14).

Brugada syndrome treatment depends on the risk of an abnormal heartbeat (arrhythmia). The patients considered at a high risk have:

- A personal history of serious heart rhythm problems
- A personal history of fainting spells

- A personal history of survived sudden cardiac arrest

Treatment may include application of an implantable cardioverter-defibrillator in persons who are at high-risk. The cardioverter defibrillator is a medical device applied on patients' chest and is connected by electrodes located in the heart. It continuously monitors the heart rhythm and if is necessary delivers electrical shocks when abnormal heartbeats (especially ventricular tachycardia or fibrillation) occurs (14).

Beta blockers are effective for patients with unstable ventricular arrhythmias. The role of Quinidine is reported to have an effect on decreasing of ventricular fibrillation in these patients. Recommendation for treatment of asymptomatic patients is not established yet. Careful observation and performing electrophysiological study in a patient from risk family is recommended in most papers in the literature (13).

CONCLUSION

Brugada syndrome is very rare arrhythmia disorder. The physicians must think of it when repeated syncope and/or nightmare were found in a healthy children and young male persons, and try to find the typical ECG findings. Rapid diagnosis and application of intra-cardiac cardioverter-defibrillator will save the life of the patient.

Conflicts of interest

The authors declare that there are no conflicts of interest.

Abbreviations:

ECG — Electrocardiography

RBBB — right bundle branch block

AV — atrioventricular conduction

EF — ejection fraction

FS — fraction shortening

CK-MB — cardiac enzyme fraction in serum

J point — repolarization phase starts at the junction or j point

ST segment — isoelectric section of the ECG between the end of S wave and beginning of the T wave. It represents the interval between ventricular depolarization and repolarization and it is sign of myocardial ischemia

ICD — intra-cardiac cardioverter defibrillator

SUNDS — sudden unexpected nocturnal death syndrome

SIDS — sudden infant death syndrome

Sažetak**BRUGADA SINDROM — PRIKAZ SLUČAJA**

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Brugada sindrom je vrsta aritmije, koja se karakteriše abnormalnim nalazom elektro-kardiograma (EKG) i povećanim rizikom od iznenadne srčane smrti. Najčešći znak na elektrokardiogramu je perzistentna elevacija ST segmenta u V1-V3 odvodima, sa blokom desne grane (RBBB). Mi predstavljamo slučaj dvanaestogodišnjeg zdravog deteta, bez ikakvih komplikacija do tada. Imao je dve epizode kolapsa/sinkope, koje su trajale dugo i spontano iščezle. Kolapsi su bili izazvani fizičkom aktivnošću. Na EKG-u se video sinusni ritam od 62 bpm, RBBB i znaci Brugade u V2 i V3 odvodima, ST elevacija ≥ 2 mm. Dete je poslato u elektrofiziološki centar u inostranstvu gde je izvršeno elektrofizi-

ološko ispitivanje. Nije pronađen nijedan akcesorni put. Atrioventrikularna provodljivost je bila normalna. Dugotrajna polimorfna ventrikularna tahikardija/fibrilacija je indukovana programiranim stimulacijama sa 3 ekstra stimulusa desnog ventrikularnog izlaznog trakta. Jednom defibrilacijom vraćen je sinusni ritam. Nakon toga su izveli EKG sa translokacijom elektroda V1-3 u drugi interkostolarni prostor i pronašli tip 1 Brugada sindroma. Nakon potvrde prisustva Brugada, tipa 1, apliciran je implantabilni kardioverter-defibrilator (ICD) na srcu deteta.

Cljučne reči: Brugada sindrom, ventrikularna fibrilacija, iznenadna srčana smrt.

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