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Concise compilation of the most clinically relevant current knowledge on Brugada Syndrome

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ABSTRACT

Brugada syndrome (BrS) is a rare disease in which arrhythmogenic changes occur in patients without any structural heart pathology. Its most dangerous complication is sudden cardiac arrest, which most often occurs around the 3rd and 4th decade of life of patients. In a similar period of time (20 - 40 years of age), the first clinical manifestations of the disease also most often occur, although this can happen even sooner in the case of a malignant form of the disease. Brugada syndrome is significantly (about 8 times) more frequently diagnosed in men, and the main factors that strongly worsen its prognosis are incidents of syncope noted in the patient's history and observed spontaneous changes in ECG studies [1][2].

KEY WORDS: Brugada syndrome, channelopathies, cardiac arrhythmias, sudden cardiac death (SCD).

INTRODUCTION

Cardiac arrhythmia is a cardiac ailment that can occur in the course of a very wide range of diseases and pathologies. It manifests itself by the presence of abnormal slowing or acceleration of the rhythm and its irregularity. The consequences of these changes can vary widely; in some cases, the course may be completely asymptomatic, but usually there will be immediate life-threatening conditions for the patient.

An example of a disease belonging to the group of pathologies that cause arrhythmia is Brugada syndrome, in

the course of which episodes of ventricular arrhythmia in the form of ventricular tachycardia and ventricular fibrillation are most frequently observed, but some patients also develop supraventricular arrhythmias, most often in the form of atrial fibrillation [3]. BrS is an autosomal dominantly inherited genetic disorder belonging to a group of so-called channelopathies, in which the gene encoding the cardiac sodium channel is mutated [4].

SYMPTOMS

The symptoms occurring in Brugada syndrome only appear during arrhythmic episodes and depend on the type and transformation of the cardiac arrhythmia currently underway. These episodes can, in some cases, be triggered by various situations, such as eating a heavy meal, the onset of fever in the patient or during sleep. The most common, and at the same time one of the least serious, symptoms are spontaneously resolving faints, which usually occurs as a consequence of rapid multiform ventricular tachycardia. Usually, the progression of prolonged ventricular tachycardia into ventricular fibrillation results in much more serious symptoms, which are sudden cardiac arrest or even death of the patient [5][6].

DIAGNOSIS

The diagnosis of Brugada syndrome is made when, in addition to the characteristic clinical signs, there are ECG changes described as type 1. These may be spontaneous or induced by the administration of antiarrhythmic drugs. These changes are ST-segment elevation ≥0.2 mV, which must be observed in one or more standard V1 or V2 leads, which may also occur in atypical leads occurring in other intercostal spaces (one or two intercostal leads above the typical site). The ST segment should then progress to a negative T-wave [7]. The diagnostic process for this condition also lists less specific ECG changes referred to as type 2 and 3. Type 2 is characterised by J-point elevation ≥0.2 mV along with saddle-shaped ST-segment elevation ≥0.1 mV and an accompanying positive T-wave, which may also be biphasic. Type 3, on the other hand, has ST-segment elevation <0.1 mV and concomitant J-point elevation ≥0.2 mV [8].

Simply observing type 2 or 3 on a patient's ECG is not in any way diagnostic, but the situation becomes different when the ECG changes to type 1 under the influence of the use of a class 1 antiarrhythmic drug in a monitored patient.

The above-described changes occurring in the consecutive 3 types of electrocardiography are presented as follows.

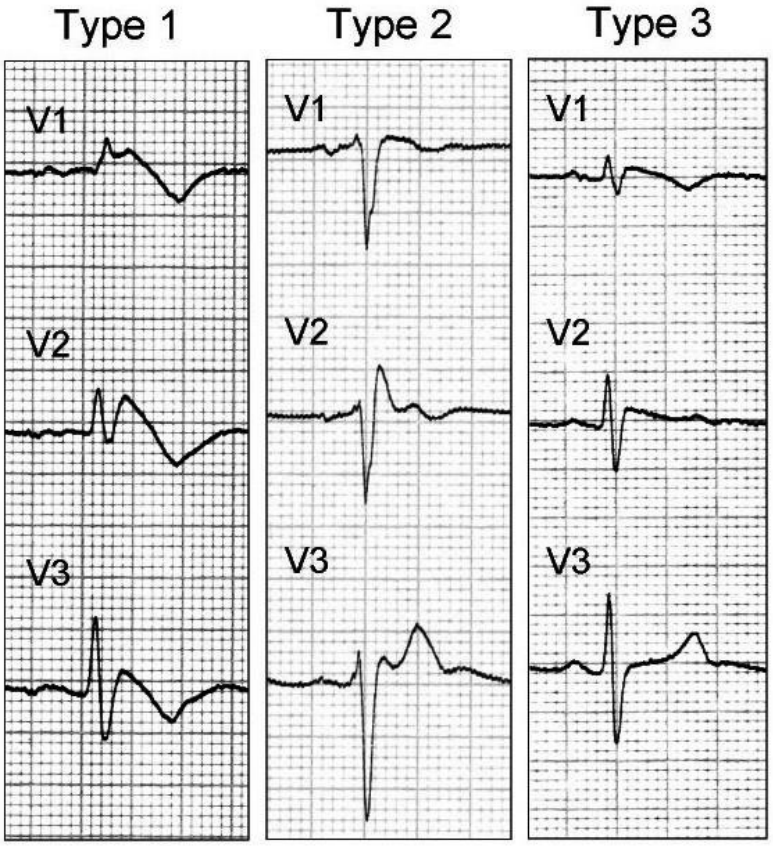

Figure 1. Changes occurring in the consecutive 3 types of electrocardiography. [9]

In a properly performed complete diagnostic process, the diagnosis of Brugada syndrome should be preceded by its differentiation from the potential presence of other diseases involving ion channels that include, for example, short QT syndrome or catecholamine-dependent multiple ventricular tachycardia [10].

TREATMENT

Treatment of patients with BrS begins with the introduction of recommendations aimed at minimising situations that may provoke arrhythmic episodes. These include avoidance of any medication that may lead to ST-segment elevation in the right atrial leads, avoidance of large meals and excessive alcohol consumption, abstinence from cocaine and, if the patient develops a fever, its rapid control with antipyretics.

In the context of pharmacotherapy, apart from the use in some cases of quinidine at a dose of 300-600 mg/d, which has the most beneficial effect, no reliable drugs are mentioned that can prevent serious consequences as a result of arrhythmia [11][12]. Its use is particularly indicated in the absence of a cardioverter-defibrillator (ICD) implantation, when interventions or supraventricular arrhythmias occur after implantation of this device.

However, the most important element of therapy is the implantation of the ICD, which is performed in patients with a documented history of cardiac arrest. However, this procedure is also performed if the patient has a history of inducible ventricular fibrillation during programmed pacing, documented ventricular tachyarrhythmias accompanied by faintness and type 1 ST-segment elevations on the patient's ECG [13][14].

A particular situation that can occur in patients is the appearance of an electron storm. This phenomenon is described as having the need for electrotherapy at least three times in one 24-hour period due to a significant increase in ventricular arrhythmias [15]. In this situation, pharmacotherapy in the form of increasing doses of

isoprenaline, starting at 0.15-0.3 μg/min, may be helpful.

For patients who, despite pharmacological treatment and having an ICD with documented recurrent interventions, an ablation procedure is a reasonable treatment alternative. It may lead to a reduction in arrhythmic episodes as a result of eliminating the sites of premature ventricular contractions leading to tachycardia or ventricular fibrillation, or removing the pericardial source of arrhythmia occurring in the right ventricular outflow tract [16][17].

SUMMARY

Brugada syndrome is a highly problematic disease mainly because its first clinical manifestation may be sudden cardiac death, without any pre-existing symptoms. Due to the rarity of the disease, it can sometimes be overlooked in the diagnostic process of cardiac arrhythmias. This situation could be minimised by increasing knowledge of BrS among clinicians. Even before an ECG is performed on patients, characteristic elements of the patient's history, such as repeated faintings in specific situations, combined with a family history of sudden deaths at a young age, can already point to the possibility of this disease. In addition, further development of scales taking into account an increasing number of relevant parameters to identify patients with the highest risk of cardiac incidents as accurately as possible seems very promising, and further development of molecular studies and knowledge of the genetic basis of BrS offers the chance to diagnose the disease more and more quickly and to guide patients through it in the best possible way [3][18][19].

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