Folia Cardiologica 2022
vol. 17, no. 2, pages 127–128
DOI: 10.5603/FC.2022.0018

ISSN 2353–7752
e-ISSN 2353–7760

Commentary to case report "Brugada syndrome: rare so, beware!" by Paweł Maciński et al.

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Brugada syndrome (BS) is a genetic disorder of ion channels resulting in their dysfunction that may lead to induction of malignant ventricular arrhythmia and sudden cardiac death [1]. The disease is diagnosed based on typical QRS complex morphology in the precordial leads in resting conditions (spontaneous type 1 Brugada pattern) or during provocative testing, although more complex definitions of BS have also been proposed [1, 2].

Multiple controversies have been related to BS, from the underlying pathophysiology through stratification of sudden cardiac death risk (including the role of programmed ventricular stimulation and electrophysiologic study [EPS]) to optimal therapy, currently with an implantable cardioverter-defibrillator (ICD) [1, 3–5]. Patient selection for ICD therapy is based on individual risk stratification but the latter remains challenging, as evidenced by the sheer number of risk factors and available prediction models. In case of a documented cardiac arrest in a patient with diagnosed BS, the situation is straightforward, as ICD is the treatment of choice for secondary prevention of sudden cardiac death [1]. In other cases, the decision may be difficult. Both the European Society of Cardiology (ESC) guidelines and two well-documented risk scores for BS (Sieira score, BRUGADA-RISK) highlight the presence of spontaneous type 1 electrocardiographic (ECG) pattern and syncope of probable arrhythmic aetiology as risk factors for future arrhythmic events [1, 5, 6]. In contrast, asymptomatic patients without spontaneous type 1 ECG pattern are usually in the "grey zone". The score developed by Sieira et al. [5] includes spontaneous type 1 ECG pattern, family history of early cardiac death (both scoring 1 point), induction of ventricular arrhythmia during EPS, syncope (both scoring 2 points), sinus node dysfunction (3 points), and aborted sudden cardiac death (4 points). Scores ≥2 were associated with a significantly higher risk of an arrhythmic event, and the risk increased with the overall score [5]. The BRUGADA-RISK model is based on somewhat different variables and includes typical spontaneous type 1 ECG pattern, spontaneous type 1 ECG pattern in the limb leads, early repolarization pattern on ECG, and syncope of probable arrhythmic aetiology. Various combinations of these risk factors determine the individual patient risk profile but each of these variables is associated with an increased risk of an arrhythmic event [6]. Clinical scores are not ideal. A validation attempt of the Sieira score by another group showed that as many as 75% of patients with an arrhythmic event in that population would not receive an ICD based on risk stratification using the Sieira score [4, 5].

Due to a reiterating role of syncope as a prognostic factor in BS, efforts should be made to evaluate the precise nature of syncopal events and exclude potential non-arrhythmic causes, such as reflex syncope, hypotension or atrioventricular block. In selected cases, implantation of an event recorder may clarify the situation and protect the patient from unnecessary ICD implantation [7, 8].

The value of EPS in the stratification of arrhythmic risk in BS is unclear [1, 5, 6, 9]. According to the ESC guidelines, induction of ventricular arrhythmia during EPS may be one factor to be considered when selecting patients for ICD therapy. In the Sieira score, 2 points are assigned for a positive EPS result, thus placing the patient at an increased risk. The BRUGADA-RISK score does not include EPS, as induction of ventricular arrhythmia was not shown to be a significant prognostic factor even in univariate analysis. It seems, however, that this diagnostic tool should not be discarded completely. In an analysis of data on 1312 patients from 8 studies, an association was found between arrhythmia induction during EPS and a history of an arrhythmic event, but the predictive value of EPS was strongly

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related to typical risk factors in BS: spontaneous type 1 ECG pattern and a history of syncope. It seems that the risk of arrhythmic events in patients without these risk factors and a negative EPS result is minimal [9].
Appropriate patient selection for ICD therapy protects high-risk individuals from an arrhythmic death, while those who do not require an ICD may be spared major complications of device therapy such as death, infective endocarditis, infection of the device pocket, inappropriate interventions, depression etc. [1, 10].

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