Letter to the Editor

What can be done when asymptomatic patients discover they have Brugada syndrome? A case report of Brugada syndrome

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Brugada syndrome is an inherited cardiac disorder associated with a specific electrocardiographic pattern, involving ST segment elevation in leads V1 to V3. When not spontaneously terminated, it can lead to ventricular fibrillation and sudden death. We present a case report of a young male whose brother suffered a sudden cardiac arrest while playing soccer. A novel mutation c.2678G>A was detected on the gene SCN5A through molecular diagnosis. The mutation was shown to be present in the individual, his daughter and his other brother. For patients with previous ventricular fibrillation and/or syncope, implantable cardiac device (ICD) is recommended. However, how can patients without symptoms but with a clear diagnosis prevent cardiac arrest?

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There are no proven prognostic markers for asymptomatic patients, and therapeutic options should be individually evaluated. Febrile state was considered to be a cause of ventricular arrhythmia, as well as sports, alcohol, drugs, electrolyte disturbances, class I anti-arrhythmic medications, and other non cardiac medications and hormones [3].

Although the occurrence of polymorphic ventricular arrhythmias is predominant during rest or sleep, there are several cases of cardiac episodes in athletes during work-out or exercise. There are no conclusive studies regarding how the physical activity may trigger inactivation of the sodium channel, and until it can be elucidated, the practice should be carefully monitored.

Febrile state is a provoking factor for malignant arrhythmia. Furthermore, a local inflammatory process was associated with cardiac disturbance. The patients’ general state of health while fever is present must be closely monitored, due to the possible induction of polymorphic tachycardia and ventricular fibrillation.

Implantable cardiac device (ICD) treatment is generally recommended for patients at higher risk of cardiac arrest, due to its cost as well as the inappropriate shocks that may occur. This treatment terminates ventricular arrhythmias, however, does not prevent future episodes. Only anti-arrhythmia drugs are indicated as a preventive treatment [4].

The role of these preventive drugs is to restore normal doming of the action potential, reducing the Ito current and increasing the calcium current. The first drug of choice would be quinidine, which prevents polymorphic ventricular tachycardia. Quinidine is a Class IA anti-arrhythmic drug that restores electrical homogeneity across ventricular myocardium and eliminates arrhythmia by phase 2 re-entry. It is also recommended as adjunct therapy with ICD and can reduce the number of shock cases. Isoproterenol (adrenergic agent) increases calcium current and is the first choice for electrical storm suppression associated with BS. Cilostazol, sodalol and mexiletine have been successful in some cases. Tedisamil is an experimental brand cardiac agent that blocks the K+ repolarization of cardiac cells [2,4].

A cardio selective and Ito specific blocker would be an ideal drug; however, there are none commercially available.

There is a need of further studies regarding the prevalence of BS cases in the same family, the related symptoms, and risk stratification. A better understanding of how these prognostic markers may trigger cardiac arrest could define what to avoid for preventive management. Genetic tests are highly recommended, because the type of mutation may determine the severity of the phenotype. There have been published cases that lead to an autosomal dominant pattern inheritance, however, the family reported in this article may be either recessive or have reduced penetrance.

Subjects carrying a mutation that promotes the premature truncation of the protein are more prone to present syncope and develop a more severe phenotype than those with a missense mutation [5]. There are many studies in asymptomatic patients, but the treatments (either drug therapy or ICD implantation) have not been clearly defined as to which would have a better cost–benefit for these patients. Many patients opt for an ICD implant as a precaution of a cardiac event; however, considering the potential flaws of the device, we question whether or not these patients will have a better quality of life.

The analysis of the family data was approved by the Ethical Committee in Research at PUCPR, protocol 808/07.

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The authors of this manuscript have certified that they comply with the Principles of Ethical Publishing in the International Journal of Cardiology [6].

References