Debate on the usefulness of ventricular stimulation for risk stratification in Brugada syndrome

La controversia de la utilidad de la estimulación ventricular para la estratificación de riesgo en el síndrome de Brugada

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The article by Dorantes and Trung, published in this issue of the journal CorSalud, provides an excellent review of a topic which, as they say, is still a matter of great controversy. It is worthwhile to do a brief historical commentary to properly situate the reader in this debate.

The now called Brugada syndrome was described in 1992 by two of the three Brugada brothers, Pedro and Josep. They managed to gather a series of 8 cases which, in their analysis, shared three characteristics: right bundle branch block, ST segment elevation and sudden death. Interestingly, the syndrome has been gradually defined along all these years. We now know, for example, that not all cases are associated with sudden death (the so-called “asymptomatic Brugada syndrome patients”), and that not all cases with the characteristic electrocardiographic pattern actually have the disease.

Establishing a differential diagnosis was from the beginning an important part of the diagnosis; however, so many cases simulating the electrocardiographic pattern of Brugada syndrome have been found now that the term “Brugada phenocopy” has been coined to distinguish them correctly. This term may well replace “pseudo-Brugada” or “Brugada-like”. It is also significant that not all cases have a true right bundle branch block but in most cases it is actually an image of a right bundle branch block. On the other hand, and not less important, the initial statement by Pedro and Josep Brugada that there was no underlying structural abnormality has crumbled due to the publication of several cases where underlying anatomical abnormalities have been demonstrated; in fact, one of the first cases of the disease, diagnosed by Dr. Pedro Brugada, was sent to Dr. Guy Fontaine and he found evidence of intramyocardial fatty infiltration.
To be fair, it is necessary to note that the Brugada brothers were not the first to describe this electrocardiographic pattern and its association with sudden death. In 1988 and 1989, the group of the University of Padua had already published two articles, in a French journal and an American journal, where they reported cases with electrocardiographic patterns that we now know are of the Brugada syndrome. They noted this fact in a letter to the editor of the Journal of the American College of Cardiology (JACC)\(^1\). In fact, the group of Dr. Andrea Nava and Dr. Bortolo Martini has made very important contributions to this condition, which can be found in detail in a chapter of a recent book on sudden death\(^11\). For several reasons, including their insistence that there were underlying structural alterations, and due to the fact that they had classified it as arrhythmogenic right ventricular dysplasia, the work of the Italian doctors did not receive the same attention as the publication of the Brugada brothers. Thanks to the Brugada brothers, many other doctors and researchers were also able to identify patients with the same characteristics and there was a sudden increase in the number of reports regarding this disease. Today, after 23 years, a search in Google with the words “Brugada syndrome” yields more than 300 000 results, while a search in PubMed/MEDLINE reports 2 599 scientific articles.

Let us go into the subject matter now. The question to answer is: what is the role of ventricular electrical stimulation in deciding the implantation of a defibrillator in Brugada syndrome? Again, the historical context is important. Ventricular electrical stimulation in Brugada syndrome began when we did not have many details about the prognosis of the disease. It began to be used much like it was used before in ischemic heart disease and other diseases associated with malignant ventricular arrhythmias\(^12\). The purpose was also to try to identify those subjects who were at risk of sudden death, taking into account whether they develop ventricular arrhythmias during ventricular stimulation; therefore, they could benefit from an implantable cardioverter defibrillator. As Dorantes and Trung\(^1\) pointed out, the initial series was given by the discoverers of the syndrome, the Brugada doctors, who showed apparently useful results of that programmed electrical stimulation for risk stratification in this disease. However, results from other groups were gradually emerging, contradicting what the Brugada brothers reported. They noted that the predictive value of ventricular electrical stimulation was actually very poor, and that it was not useful to stratify the risk of sudden death. This is the origin of the controversy, which persists to the present time. However, after the conduction of several studies by various groups other than the Brugada brothers, which were very well analyzed and presented by Dorantes and Trung\(^1\), it is possible to state that programmed ventricular stimulation does not have a significant role in risk stratification for Brugada syndrome.

This assertion is supported by the latest report by experts from the Heart Rhythm Society\(^13\). As they rightly point out, if the patient already had an episode of sudden death or resuscitated cardiac arrest, a defibrillator must be implanted; on the other hand, if the patient is asymptomatic, the risk is so low that it is not necessary to conduct an electrophysiological study, nor implant a defibrillator. The authors analyze and detail the results of programmed electrical stimulation for other diseases associated with sudden death that had previously been classified as idiopathic or primary ventricular fibrillation. The analysis shows that Brugada syndrome is an entity. The experience of the authors is clear on this. They also point out clearly why we must be careful in indicating a defibrillator, especially with regard to possible complications. In this sense it is important to consider that many patients with Brugada syndrome are young men who will require multiple devices throughout their life, which substantially increases the risk of these complications, not only in relation to the implant but also regarding the psychological consequences of both appropriate and inappropriate electric shocks. The usefulness of quinidina\(^14,15\) and the problem of its scarcity in many countries\(^16\) are treated clearly by the authors.

We would like to finish this Editorial making mention of a forthcoming book by the Dominican Society of Cardiology with the backing of the Inter-American Society of Cardiology\(^17\). It is a monograph on sudden death which addresses in detail this major public health issue, and it includes, of course, Brugada syndrome.

REFERENCES
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