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EDITORIAL COMMENT

Pediatric Brugada Syndrome

Avoiding the Inappropriate ICD*

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Brugada syndrome (BrS) is an inherited ion channelopathy characterized by a coved-type ST-segment in the right precordial leads predisposing to ventricular arrhythmias and sudden death (1). Clinical expressivity varies ranging from a complete lack of symptoms in 72% of patients to sudden death as an initial presentation in 7% (2). Follow-up results from the earliest patient experience exposed dismal results wherein 62% of patients who presented with sudden cardiac death had recurrence of ventricular arrhythmias (3).

With the 25th anniversary of the original description of BrS upon us, it is appropriate to revisit progress made, particularly as it pertains to patient outcomes and the prevention of sudden cardiac death. Despite significant advances in our understanding of BrS from a cellular and mechanistic standpoint, as practitioners we continue to struggle with identifying the asymptomatic patient at risk for sudden cardiac death, specifically as it pertains to primary prevention implantable cardioverter-defibrillator (ICD). The case for risk stratification, particularly in the pediatric patient, is demonstrated by the fact that most pediatric patients diagnosed with BrS are asymptomatic at presentation. In a study by Harris et al. (4), the most common initial presentation of BrS was a family history (47%), followed by an incidental electrocardiographic finding (25%), syncope (14%), arrhythmia (13%), and aborted sudden cardiac death (1%); therefore, 72% of pediatric patients diagnosed with BrS were asymptomatic at presentation.

As with many cardiovascular diseases in the young, the paucity of robust clinical research has forced the pediatric electrophysiologist to extrapolate from adult data. Our struggle with the rarity of BrS in pediatrics is found in 1 of the first pediatric-specific papers addressing this population. In a study by Probst et al. (5), only 30 pediatric patients with BrS were identified in 13 tertiary institutions over a 15-year follow-up period. We have therefore been limited in making sound clinical decisions that open the door for inappropriate, and rather burdensome, therapy.

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In this issue of the *Journal*, Gonzalez Corcia et al. (6) present long-term outcomes of pediatric patients implanted with ICDs. The study details a single institution experience of 35 patients, the largest of its kind, wherein ICDs were implanted at a mean age of 13.9 years over a follow-up period of 7.3 years. The study reflects on many important points, not the least of which is the authors' judicious assessment of risk. Since the study's inception in 1992, coincident with the first published description of the disease, 210 patients (<20 years of age) in the 24-year study period were diagnosed with BrS. Among the patients identified, only 16% went on to ICD implantation. Of those 35 patients undergoing implantation, only 3 (8%) were asymptomatic; however, all 3 of these patients had a spontaneous type I electrocardiographic pattern. Compare this study's implant proportion of 16% with the recently published data on an adult cohort recently in the *Journal* of 28% (7). Although ICD therapy has been clearly shown to prevent sudden cardiac death, only a small minority of patients deemed by these expert authors as "at risk" met criteria for implantation. As pediatric electrophysiologists, it is important to keep these proportions under consideration when we reflect on the treatment of our own BrS patients. If 1 is substantially exceeding the 16% implant proportion in this article in their own BrS population, prompt reconsideration of institutional risk stratification algorithms should ensue.

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Determining risk for sudden cardiac death lies in examining the event rate in this study population. Among the 8 (23%) patients receiving appropriate ICD shocks and the 1 (3%) with successful anti-tachycardia pacing for ventricular arrhythmias, all were men and presented with symptoms before device implantation. Not surprisingly, aborted sudden cardiac death was more predictive of receiving an appropriate shock than was a presentation of syncope. Of the patients with a spontaneous type I BrS pattern on electrocardiography, 40% received appropriate therapy. Most importantly, no asymptomatic patients presented with an appropriate shock. Similar to previously published adult studies, symptoms and a type I BrS pattern predicted appropriate ICD therapy (7,8).

The value of programmed ventricular stimulation (VPS) as a predictor of sudden death has been controversial (7,9), and pediatric reports have been scarce (2). The authors in this study did not find predictability in VPS. This does not imply, however, that VPS carries no value. Perhaps the utility in VPS is identifying other substrates for arrhythmia, whether that be conduction abnormalities, atrial standstill, or supraventricular tachyarrhythmias. Identifying these substrates before ICD therapy is reasonable, such that well-defined programming parameters can be instituted with the objective of reducing inappropriate therapy.

Unfortunately, device complications continue to sober optimism in pediatric ICD literature and the data presented in this paper are no different (10,11). This study reports a 20% inappropriate shock rate caused by supraventricular tachycardia, lead fracture, and T wave oversensing. Five (14%) patients experienced device related complications resulting in immediate device revision. Although device complications trended downward after 2010 in this study, pediatric electrophysiologists must have candid conversations with patients and families regarding these complications. Instead of emphasizing the indications for implantation, as practitioners we must emphasize the indications to exercise caution and even humility. In the current state of device technology, it is inappropriate and unethical to implant a device without detailing the potential for adverse outcomes. The expectation should be that appropriate and inappropriate shocks, as demonstrated in this paper and others, are of near equal probability, even in carefully selected ("at risk") patients. This is no more evident that in the case of pediatric BrS.

Despite the fact that ICD therapy is the only proven intervention to prevent sudden cardiac death, overzealous implantation should be avoided at all costs. In the current state of device technology, even in carefully selected at risk patients as described in this study, the dark side of implantation is unavoidable. The decision to implant an ICD in a pediatric patient is a team sport, consisting of the implanting physician/surgeon, the pediatric patient, and the family. Only programs that are equipped to provide pediatric-specific social support and mental health care are appropriate to provide this subspecialized treatment modality. The implanter is merely the facilitator of these conversations. The authors of this work should be applauded for focusing on the often forgotten pediatric patient and providing practitioners data to avoid the inappropriate implant.

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