Brugada Syndrome in Children
Don’t Ask, Don’t Tell?

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In recent years, numerous asymptomatic individuals worldwide have undergone electrophysiological studies “only” because they have a pathological ECG indicative of Brugada syndrome. Furthermore, at least 1 of 3 of these individuals had inducible ventricular fibrillation (VF) and underwent implantation of a cardioverter defibrillator (ICD). “Asymptomatic Brugada syndrome with inducible VF” became an accepted indication for ICD implantation because data from a single large study showed that 12% of such patients develop spontaneous VF within 3 years of diagnosis. More recent (albeit smaller) studies, however, suggested that the risk for spontaneous VF in asymptomatic patients is lower. In fact, the most recent studies show that only 3% to 4% of individuals undergoing ICD implantation for “asymptomatic Brugada syndrome with inducible VF” developed spontaneous VF within 3 years of implantation. At the same time, as many as 28% of them developed ICD-related complications. The realization that we have done more harm than good to many asymptomatic individuals has reopened the debate on the optimal management of asymptomatic Brugada syndrome. Indeed, defining the role of electrophysiological testing in asymptomatic Brugada syndrome is probably the most heated debate in arrhythmology nowadays.

The article referenced in the text provides a detailed analysis of the evidence and the ethical implications of this debate. The author argues that the prevalence of spontaneous VF in asymptomatic Brugada syndrome is lower than previously thought, and that the risk of adverse events associated with ICD implantation is higher. The author suggests that the use of ICDs in asymptomatic Brugada syndrome should be reconsidered, and that other strategies, such as close monitoring and the use of medications, should be considered instead.

The article also discusses the genetic basis of Brugada syndrome, with a focus on the role of the SCN5A gene, which is mutated in most cases. The author highlights the importance of genetic testing for Brugada syndrome, especially in families with a history of the disease.

The implications of these findings are significant, as they challenge the current paradigm of managing asymptomatic Brugada syndrome. The author calls for a reevaluation of the indications for ICD implantation in this population, and for a more personalized approach to risk assessment and management.

The article is published in a reputable medical journal, and is available for further reading.

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ing.\textsuperscript{15} Although the repolarization abnormalities (explained above) facilitate the onset of polymorphic ventricular tachycardia, it is the depolarization disturbance (conduction slowing leading to wave break of the reentrant wave) that allows the ventricular tachycardia to become sustained and to degenerate to VF.\textsuperscript{16} In addition, recent data suggest that adult men with Brugada syndrome have higher levels of testosterone than age-matched healthy men,\textsuperscript{17} and it is not clear when this abnormal testosterone rise takes place.

Many electrophysiologists adopted a “don’t ask, don’t tell” policy toward the need for screening the offspring of adults with Brugada syndrome. This policy resulted from 2 factors: the perception that carriers of Brugada mutations almost never develop arrhythmias during childhood and the fact that prophylactic ICD implantation in children incurs significant morbidity. The present series of Brugada syndrome in children\textsuperscript{9} could lead to a reversal of this conservative approach. Thus, a few points are noteworthy. First, only through the collaboration of investigators from 13 different referral centers in 3 European countries was it possible to report 30 children.\textsuperscript{9} This means that during the 15 years that passed since the original description of the Brugada syndrome, <3 children (on average) were identified at each of the tertiary centers joining forces for this report. In contrast, >1500 adults with Brugada syndrome have been reported.\textsuperscript{4} Thus, this series supports, rather than contradicts, the notion that symptomatic Brugada syndrome is rare in children. Second, having a history of familial sudden death has not been shown to be of prognostic value in adults\textsuperscript{4} and does not appear to be of prognostic value in children either.\textsuperscript{3} Third, asymptomatic adults who have a type I Brugada ECG only when challenged with sodium-channel blockers are at low risk for arrhythmic events (during follow-up periods of ≈3 years) even if they have inducible VF.\textsuperscript{4} In the present series, only 1 of 13 children (8%) who had their Brugada ECG unraveled by drug challenge actually developed symptoms. Thus, although the drug challenge test appeared to be safe in children,\textsuperscript{9} one should carefully consider whether the limited new information derived from such test merits the anxiety resulting from a positive test in a child. Finally, quinidine is extremely effective for preventing inducible VF in the electrophysiological laboratory\textsuperscript{18} and appears to be effective for preventing spontaneous arrhythmias in adults with symptomatic\textsuperscript{21} and asymptomatic\textsuperscript{18,19} Brugada syndrome. The few children who received quinidine in this series did well.\textsuperscript{9}

How should we implement these new data when advising parents with Brugada syndrome? First, we ought to acknowledge that we do not know enough about prognosis or risk stratification of Brugada syndrome in children. Because of our limited knowledge, some parents may prefer not to know if their asymptomatic children have a Brugada-type ECG, and we ought to respect such decision. Other parents may take the opposite attitude, insisting on doing “whatever it takes” to prevent arrhythmic death. In practical terms, however, doing “everything possible” to prevent sudden death starts with ECG recording but may culminate in prophylactic ICD implantation. It is imperative that such parents comprehend that the alarmingly high rate of ICD-related complications reported for young adults with Brugada syndrome\textsuperscript{6} is likely to be worse among children. Because of their smaller vessel diameter and cardiac size, infants with intravenous defibrillator leads often develop venous obstruction and tricuspid regurgitation,\textsuperscript{20} whereas epicardial defibrillator patches require extensive surgery and are associated with high defibrillation thresholds.\textsuperscript{20} Moreover, disruption of the fixed leads eventually occurs as children grow.\textsuperscript{20} Although novel ICD implantation techniques are being developed,\textsuperscript{21} the need for repeated surgical revision continues to be a serious problem. Totally extracardiac ICD systems delivering only shock therapy will soon be available, and children with Brugada syndrome may end up deriving the greatest benefit from such systems. In the meantime, relatives of patients with Brugada syndrome should be taught the art of cardiopulmonary resuscitation and be informed of the availability of external automatic defibrillators. In addition, because fever is an important trigger for arrhythmias in Brugada syndrome, it makes sense to use antipyretics liberally and to hospitalize children during febrile episodes,\textsuperscript{9} particularly when fever causes additional ST-segment elevation. Finally, the possibility of drug therapy should be discussed. Because sentences like “the ICD is the only effective therapy for Brugada syndrome” appear in every review of the topic, many physicians may not even consider drug therapy. One should recall, however, that children with congenital long-QT syndrome have received drug therapy with β-blockers for many years, and in large patient subgroups, this therapy proved to be effective in preventing arrhythmic death. Ironically, many of these now youngsters with congenital long-QT syndrome were “saved from the ICD” simply because these devices were not available when the long-QT syndrome was described. Because of its strong I\textsubscript{Na}-blocking capabilities,\textsuperscript{22} its efficacy in preventing inducible and spontaneous VF,\textsuperscript{18,19} and its efficacy in terminating VF storms,\textsuperscript{23} quinidine therapy has been proposed as a “bridge to ICD” for symptomatic children with Brugada syndrome\textsuperscript{23} (to prevent VF recurrence until infants grow sufficiently to undergo ICD implantation safely). It is too early to recommend universal prophylactic quinidine therapy for asymptomatic children with Brugada syndrome, but for parents and physicians willing to adopt an active approach, it is an option to consider.

Disclosures

None.

References


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