Editorial Comment

Is the Implantable Defibrillator Indicated in Patients With Hypertrophic Cardiomyopathy and Aborted Sudden Death?*

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The natural history of patients with hypertrophic cardiomyopathy (HCM) is characterized by slow progression of symptoms (i.e., angina or dyspnea, or both) and left ventricular hypertrophy. Severe functional limitation is unusual and is encountered in only ~20% of patients. Approximately 50% of deaths are sudden (1). Although complete heart block may complicate myocardy, the development of symptomatic conduction disease in patients not undergoing operation is rare. Preexcitation syndromes may be present in a small proportion of patients (2%) but appear to be seldom responsible for sudden death in HCM. A short PR interval with a broad QRS complex might mimic the presence of a Wolff-Parkinson-White syndrome in some patients with HCM. This finding may be explained by an abnormal septal activation of the hypertrophied septum. We recently studied a cohort of 65 patients (unpublished observations) with HCM and either documented or suspected ventricular tachyarrhythmias undergoing invasive electrophysiologic study. Surprisingly, in ~20% of patients, enhanced atrioventricular (AV) node conduction was observed, allowing 1:1 conduction with ventricular rates up to 280 beats/min in the absence of an accessory pathway. This finding may be of clinical importance as a possible trigger mechanism of sudden arrhythmic death in HCM because these high ventricular rates during paroxysmal atrial fibrillation may be detrimental in patients with HCM and impaired diastolic filling. Outflow tract obstruction and coronary spasm have been suggested as a causal factor. However, these mechanisms have not yet been shown to be of importance. Further hypotheses, currently also not proven, have implicated acute changes in diastolic filling, acute myocardial ischemia or impaired autonomic regulation as causes of sudden death. Currently, not much information is available concerning the mechanism of sudden death in HCM, making risk stratification difficult.

Presently, no convincing data are available for effective risk stratification of patients with HCM (2–4). This inability to correctly identify patients at risk of sudden cardiac death in HCM reflects our poor understanding of the mechanisms of sudden death in this entity. Although some initiating mechanisms have been identified in single cases, such as atrial fibrillation with rapid ventricular conduction, altered baroreflex control of peripheral blood flow and others, the extent of myocardial disarray and a potential for ischemic events in patients with HCM have to be better characterized before appropriate pharmacologic or nonpharmacologic interventions, such as myectomy or implantation of an implantable cardioverter-defibrillator (ICD), are undertaken.

The available data suggest that ventricular tachyarrhythmias are the cause of sudden death in most patients with HCM, either as a primary event related to an arrhythmogenic substrate or as a secondary phenomenon triggered by myocardial ischemia, diastolic dysfunction, outflow tract obstruction or supraventricular tachyarrhythmias. Conventional risk factor stratification, including family history, presence of syncope and nonsustained ventricular tachycardia (VT) during Holter monitoring, identifies a cohort of patients at increased risk for sudden cardiac death. In ~30% of this group a probable initiating mechanism that is amenable to specific therapy can be identified. Treatment may be targeted at the prevention of paroxysmal atrial fibrillation, which can be effectively prevented with amiodarone; conduction disease by a cardiac pacemaker; rapid AV conduction through an accessory pathway by radiofrequency catheter ablation; ischemia with high dose verapamil; and left ventricular outflow tract obstruction by myectomy or dual-chamber pacing or, recently, by occlusion of the septal branches of the left anterior descending coronary artery. However, in the remaining group (~70%), the patient is recognized as being at increased risk, but there are either multiple potential triggers or nonidentifiable triggers that can be targeted. There is general agreement that patients with cardiac arrest and documented ventricular fibrillation, who rarely survive this event, or patients with HCM and episodes of sustained monomorphic VT, although rare in HCM, represent the patient cohort at highest risk for sudden death.

In the present issue of the Journal, Primo et al. (5) report the results of a retrospective analysis of the occurrence of cardiac events during follow-up in 13 patients with HCM who received an ICD because of either aborted sudden death (n = 10) or sustained ventricular tachycardia (n = 3). The results and findings were compared with those in 215 patients with an ICD and other structural heart diseases or idiopathic ventricular fibrillation. During the mean follow-up period of 26 ± 18 months, only 2 of 13 patients with HCM received appropriate shocks. The calculated cumulative incidence of shocks was 21% in the patients with HCM and 66% in the remaining patients after 40 months. The patient cohort with HCM is

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poorly characterized concerning clinical characteristics and echocardiographic findings as well as the circumstances of the cardiac event before ICD implantation. Seven of 13 patients received amiodarone before ICD implantation for unspecified reasons. After ICD implantation, all but one patient received either a beta-adrenergic blocking agent, verapamil or amiodarone, making assessment of the impact of ICD implantation on prognosis impossible. The conclusion that the ICD seems to have a less important impact on prognosis in patients with HCM than in patients with other etiologies of aborted sudden death is poorly substantiated by the authors. Additionally, the report compares a small patient cohort with a large patient group with different underlying cardiac etiologies. Given the small patient numbers and the varying follow-up period in the small group of patients with HCM, the authors should have given standard deviations in the Kaplan-Meyer event curves to illustrate that their observations were due to chance in a highly selected group of patients with HCM.

Although the use of an ICD remains controversial in patients with HCM, and there is a lack of information in the published reports, three recent studies, presented only in part at meetings, have addressed the outcome in patients with HCM and an ICD. Silka et al. (6) reported on 44 patients with HCM of a cohort of 177 young patients (<20 years old) receiving an ICD. In the subgroup of 44 patients with HCM, 25 (57%) received appropriate ICD discharges during a mean follow-up period of 31 ± 23 months. A similar observation was made by Tripodi et al. (7) who reported on 31 patients with an ICD and HCM who were followed up over 33 ± 7 months, with a reported incidence of 32% appropriate discharges. At our referral center, <2% of ICD recipients were diagnosed as having HCM. Fourteen patients received an ICD and were followed up for 48 ± 24 months; 43% of patients had an appropriate ICD intervention during follow-up, as assessed by third-generation devices with memory function.

Taking these observations into account in comparing the low event rate in the present study by Primo et al. (8), one is left with the impression that one is dealing with a highly selected subgroup of patients in which additional pharmacologic therapy may have had some impact on the low event rate during follow-up. This controversy underlines the need for a prospective trial in high risk patients with HCM and the need for a registry to collect data on the outcome of patients undergoing different therapeutic interventions, such as DDD pacing, septal ablation, myectomy or ICD implantation, alone or in combination.

Currently, in Europe there is a prospective registry on HCM and ICD implantation on the way supported by the Working Group on Arrhythmias and the Working Group on Myocardial and Pericardial Diseases of the European Society of Cardiology, which was initiated by Martin Borggrefe, Münster, Germany and William McKenna, London, United Kingdom. Possibly, this registry will give some definite answer concerning the role of ICD implantation in cardiac arrest survivors and HCM. Whether other nonpharmacologic interventions, such as septal ablation, DDD pacing or myectomy, have some impact on the management of sudden cardiac death survivors remains speculative.

Recently, we prospectively studied 10 patients after cardiac arrest who underwent myectomy for hypertrophic obstructive cardiomyopathy (HOCM) (8). Eight of 10 patients had inducible sustained ventricular tachycardia or fibrillation before operation. After myectomy, only no to six ventricular responses were inducible in all 10 patients by means of a stimulation protocol that included up to three extrastimuli. No recurrent arrhythmic event occurred during a follow-up period of 4.5 years in the absence of antiarrhythmic drugs. This preliminary observation may be explained by surgical removal of an “arrhythmogenic substrate” in some patients with HOCM and life-threatening tachyarrhythmias and may support a potential mechanism for the beneficial long-term results after surgical treatment. Therefore, in patients with cardiac arrest and inducible ventricular tachyarrhythmias, myectomy may be the treatment of choice and presents a curative approach in contrast to implantation of an ICD.

Conclusions. The report by Primo et al. (5) initiates a new discussion on the value of an ICD in patients with HCM and aborted sudden cardiac death. At present, the patient with sustained ventricular tachyarrhythmias and HCM should undergo implantation of an ICD (9), unless there is a specific trigger that can be effectively targeted or a potential arrhythmogenic substrate that can be abolished effectively by myectomy, especially in patients with HOCM or apical aneurysms. The low peri-implant mortality and morbidity of the transvenous lead systems make ICD implantation a logical and cost-effective form of therapy. The availability of improved logging of arrhythmic events and the rhythm immediately preceding arrhythmia detection may provide new information in this group that in future will help to decide with more security which patient should undergo ICD implantation.

References

