Sudden Death in Myotonic Dystrophy: The Potential Role of Bundle-Branch Reentry

To the Editor:

We read with interest the recent report of Lazarus et al. This is an important study addressing the prevalence of His-Purkinje conduction disturbances and ventricular arrhythmia inducibility by a standard pacing protocol in a large sample of patients with myotonic dystrophy (MD) and asymptomatic conduction abnormalities. However, several points of the discussion should be reviewed owing to possible misinterpretation.

First, the authors found a high frequency of prolonged HV interval and raised the issue of prophylactic pacing to limit the high incidence of sudden death seen in MD. However, reports of sudden death in patients with MD after pacemaker implantation (including 1 patient in the present study) and of sudden death shortly after clinical documentation of sustained monomorphic ventricular tachycardia (VT) make VT in MD a potential cause of sudden death likely enough to reconsider the authors’ insinuation of prophylactic pacemaker implantation. Furthermore, demonstration of HV-interval prolongation should not be taken only as a risk factor of AV block, because VT in MD typically has a bundle-branch reentrant mechanism, a mechanism that is associated with slow conduction in the His-Purkinje system.

Second, the authors reported a higher inducibility of polymorphic ventricular arrhythmias in younger patients than in older patients, speculating that evolution to sudden death of patients with positive inducibility in the former group could account for the lower inducibility in the latter group. However, inducibility of polymorphic ventricular arrhythmias is often considered nonspecific, especially if induced with aggressive ventricular stimulation, as in the present study (200-ms coupled triple extrastimuli). In addition, sustained monomorphic VT is the clinical ventricular arrhythmia most frequently reported in MD. No sustained monomorphic VT was induced in the present study, which used a standard stimulation protocol. Standard protocols have proved valuable in patients with myocardial reentrant VT but often fail to induce bundle-branch reentrant VT, which may require the use of specific pacing protocols (long-short–coupled extrastimuli), infusion of drugs (isoproterenol, procainamide), or both. This factor could have accounted for the noninducibility of monomorphic VT in the present study.

The authors may like to clarify these issues by reporting on any patient with MD who developed sustained monomorphic VT that was not induced during the electrophysiological testing, such as the one already presented in the preliminary report of this work.

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Response

First of all, we would like to thank Dr Merino et al for the interest they expressed in our report, and in answer to their letter, we wish to make the following comments.

All our patients were not asymptomatic; 3 had a high degree of AV block, 12 had presyncope/syncope, and 11 experienced palpitations. However, none had documented spontaneous sustained monomorphic ventricular tachycardia (SMVT) before electrophysiological (EP) testing.

Arrhythmic sudden deaths, attributed either to ventricular arrhythmias or to AV block, have been described in myotonic dystrophy (MD). For us, having a prolonged HV interval clearly raises the problem of prophylactic pacing, because the risk of subsequent spontaneous paroxysmal high-degree AV block is then clearly higher than the risk of ventricular tachyarrhythmia. Sudden death can also be nonarrhythmic in MD, as demonstrated by typical cases of sudden death that occurred in MD patients equipped with pacemakers in whom extended diagnostic capabilities could eliminate an arrhythmic cause.

In our experience, which now includes 159 EP testing procedures in 137 myotonic patients, we never observed initiation of SMVT during programmed ventricular stimulation, although we systematically used short-long coupling extrastimuli (but without drug infusion). Concerning the inducibility of polymorphic ventricular arrhythmias, we agree that this is often considered nonspecific, but this remains to be demonstrated in the peculiar MD population in which the arrhythmic risk is different from the general population. To illustrate this, during the follow-up of our MD population, we only observed the clinical occurrence of spontaneous SMVT in 1 case, and this patient had a nonsustained polymorphic VT previously induced during EP testing. Under amiodarone treatment, VT did not recur after 35 months of follow-up.

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_Circulation_. 2000;101:e73
doi: 10.1161/01.CIR.101.5.e73
_Circulation_ is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the
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