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What is the best way to detect infra-Hisian conduction abnormalities and prevent sudden cardiac death in myotonic dystrophy?

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Myotonic dystrophy type 1 (DM1), also called Steinert's disease, is a multisystemic disorder for which the cardiologist is often consulted. Clinical manifestations of this disease, which involves multiple systems, may include muscle weakness, myotonia, multiple endocrinal disorders, cataract, respiratory insufficiency, conduction system disease, supraventricular arrhythmias, ventricular tachyarrhythmias and dilated cardiomyopathy.[1–3]

Sudden cardiac death is the second most common mode of death after respiratory failure in this population and occurs in up to one-third of patients.[4] As DM1 becomes apparent in most cases by extracardiac manifestations, neurologists usually refer patients to cardiologists in order to evaluate their risk of cardiac sudden death and to suggest the best measures to prevent it. In this case, interventional measures are part of a primary prevention strategy in patients, often with no personal history of cardiac disease, such as patients in whom hypertrophic cardiomyopathy or Brugada syndrome are diagnosed fortuitously or by systematic familial screening. Less frequently, cardiologists are the first specialists who see the patient with predominant or exclusive cardiac symptoms, illustrating the common dissociation in expression of the disease from one tissue to another.

Decision-making is complex in patients with DM1, mainly owing to the absence of strong prognostic factors for sudden death. Conduction defects on the ECG or a personal history of atrial fibrillation are the only proven prognostic factors for sudden death, but their positive predictive value was estimated at only 12%,[5] which is not a sufficient criterion to apply specific therapeutic measures in a given patient. Studies evaluating other non-invasive tools, such as late ventricular potentials, as prognostic factors were also disappointing. The difficulty in identifying prognostic factors in DM1 is also related to the involvement of multiple systems, their potential implication in sudden death and the difficulty in identifying the precise mechanisms underlying sudden death, even in patients for whom premortem cardiac rhythm is recorded.[6] Progression of conduction system disease to complete atrioventricular (AV) block has always been presumed to be the cause of sudden death in most patients, in view of its high prevalence, and prophylactic permanent pacing has been recommended by the American College of Cardiology and the American Heart Association[7] in patients with first degree AV or fascicular block on the ECG (class IIb indication) owing to the 'unpredictable evolution of the disease'. However, ventricular tachyarrhythmias and extracardiac complications, such as acute respiratory failure or pulmonary embolism, may also be implicated and may account for sudden deaths in patients with pacemakers.[6]

In this context, what is the preferred strategy for a patient to prevent sudden death? In view of the low positive predictive value of non-invasive cardiac evaluations, an electrophysiological testing-based approach has been proposed to improve patient selection for prophylactic pacing. Its objective is to obtain the most accurate assessment of conduction defects possible. The potential benefit of this invasive approach has been supported by an observational study, in which patients presenting with DM1 who had conduction abnormalities on surface ECG and an HV interval >70 ms, had a high rate of complete AV block.[6] In clinical practice, in many cardiology units in the USA and in Europe, patients in whom conduction defects are identified on the ECG undergo electrophysiological testing and prophylactic pacing when the HV interval is >70 ms, even if this strategy is not supported by studies showing a benefit for sudden death prevention.

In their paper recently published in Heart, Lallemand *et al* provide interesting information to clarify the use of this electrophysiological testing-based strategy.[8] Their study illustrate that in patients with DM1 with an initial electrophysiological study that did not fulfil criteria for prophylactic pacing, modifications on the resting ECG on annual check-up were associated with significant worsening of infra-Hisian conduction defects. These results suggest that, in clinical practice, after an initial invasive evaluation of infra-Hisian conduction, a non-invasive electrophysiological follow-up may be useful to detect the occurrence of severe infra-Hisian conduction abnormalities and could represent an indication for new electrophysiological testing.

Finally, the key point of controversy about the use of an invasive strategy based on systematic electrophysiological study and prophylactic pacing is to know whether it is associated with a significant clinical benefit for patients. To answer this question, a controlled study examining sudden death or other major cardiac events is required.

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