Supplementary Online Content

Wahbi K, Meune C, Porcher R, et al. Electrophysiological study with prophylactic pacing and survival in adults with myotonic dystrophy and conduction system disease. *JAMA*. 2012;307(12):1292-1301.

This supplementary material has been provided by the authors to give readers additional information about their work.

eResults. Analyses of overall survival in imputed datasets, overall survival in the original dataset, and sudden death in the original dataset

Analysis of overall survival in imputed datasets

The Cox model adjusted on the propensity score with the linear predictor scale, instead of the propensity score quintiles, yielded a significantly lower overall mortality in the IS than in the NIS group (HR 0.59; 95% CI 0.36 to 0.97, *P*=.036). After further adjustments for the baseline prognostic factors, HR was 0.56 (95% CI 0.36 to 0.97, *P*=.036).

Analysis of overall survival in the original dataset

Analysis of the original dataset excluding observations with missing data from the adjusted models yielded a significantly lower overall mortality in the IS than in the NIS group. The value of HR was a) 0.58 (95% CI 0.35 to 0.98, *P*=.040) after adjustment for the propensity score quintiles, b) 0.52 (95% CI 0.31 to 0.90, *P*=.018) after adjustment for the propensity score quintiles and baseline prognostic factors, c) 0.58 (95% CI 0.34 to 0.97, *P*=.037) after adjustment for the propensity score, and d) 0.56 (95% CI 0.33 to 0.95, *P*=.033) after adjustment for the propensity score and baseline prognostic factors. Furthermore, an analysis of 216 patients in the IS group matched with 108 patients in the NIS group by propensity score revealed an even more significantly lower mortality in the IS than in the NIS group (HR 0.46; 95% CI 0.27 to 0.71, *P*=0.005). After further adjustment for baseline prognostic factors, HR was 0.40 (95% CI 0.22 to 0.71, *P*=0.002).

Analysis of sudden death in the original dataset

An unadjusted group comparison in the original dataset yielded a lower incidence of sudden death in the IS than in the NIS group (HR 0.28; 95% CI 0.13 to 0.61, *P*=0.001). In the dataset including all cases, the incidence of sudden death was also lower in the IS group. The value of HR was a) 0.28 (95% CI 0.11 to 0.67, *P*=0.005) after adjustment for the baseline prognostic factors, b) 0.24 (95% CI 0.10 to 0.59, *P*=0.002) after adjustment for the propensity score quintiles, and c) 0.22 (95% CI 0.09 to 0.56, *P*=0.002) after adjustment for the propensity score quintiles and baseline prognostic factors. In the matched dataset, the value of HR was 0.21 (95% CI 0.07 to 0.63,

P=0.005) and 0.18 (95% CI 0.05 to 0.61, *P*=0.006) after adjustment for the baseline prognostic factors. These observations are concordant and support the results obtained in the imputed datasets.

eTable 1. Variables used to develop the propensity score

Age
Sex
Number of CTG repeats
Age at disease onset
Muscular disability rating score
Walton muscle weakness score
Familial history of sudden death
Diabetes
Dyslipidemia
Smoking
Coronary artery disease
History of supraventricular tachyarrhythmia
History of ventricular tachyarrhythmia
History of heart failure
History of syncope
Dyspnea
Light-headedness
Palpitations
Systolic blood pressure
Diastolic blood pressure
Heart rate
PR interval
QRS duration
Left bundle branch block
Right bundle branch block
Left ventricular ejection fraction at study entry
Vital capacity
Amiodarone at baseline
Oral anticoagulant at baseline
Aspirin at baseline
Class I antiarrhythmic drugs at baseline
Beta-adrenergic blocker at baseline
ACE inhibitors/ARB at baseline

eTable 2. Individual characteristics of the patients who died suddenly in the IS group

Pt no	Age (y) at death/sex	CTG repeats	Walton Muscle weakness score	Personal history of ventricular arrhythmia	Baseline ECG observations	Baseline EPS observations		Therapy		Sudden death information		
						HV interval (ms)	Programmed ventricular stimulation	Device	Antiarrhythmic drugs	Device memory interrogation	Cardiac rhythm	Witnessed
1	20/M	835	0	No	1st degree AV block	80	Positive	PM	-	Yes	Ventricular fibrillation	Yes
2	48/M	330	4	No	1st degree AV block	74	Positive	ICD	Amiodarone	Yes	Asystole with pacing stimuli	No
3	54/F	730	3	No	LBBB	70	Negative	PM	-	Yes	Asystole with pacing stimuli	No
4	54/M	1330	6	No	1st degree AV block	80	Negative	PM	-	No	Undetermined	No
5	52/M	660	4	Yes	1st degree AV block	80	Negative	PM	-	Yes	Asystole with pacing stimuli	Yes
6	62/F	1160	9	No	LBBB	75	Negative	PM	-	Yes	Ventricular fibrillation	Yes
7	56/F	1330	3	No	Non-specific QRS widening	62	Negative	None	-	-	Undetermined	No
8	56/M	200	3	No	1st degree AV block, RBBB	74	Negative	PM	Beta-adrenergic blocker, amiodarone	Yes	Ventricular fibrillation	No
9	72/F	80	2	No	1st degree AV block, LBBB	-	Negative	PM	-	Yes	Asystole without pacing stimuli	Yes
10	61/M	400	4	No	1st degree AV block	70	Positive	PM	-	No	Undetermined	No

AV = atrioventricular; EPS = electrophysiological study; ICD = implantable cardioverter defibrillator; LBBB = left bundle branch block; PM = pacemaker; RBBB = right bundle branch block