

## Supplementary Appendix

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

Supplement to: Groh WJ, Groh MR, Saha C, et al. Electrocardiographic abnormalities and sudden death in myotonic dystrophy type 1. *N Engl J Med* 2008;358:2688-97.

**Online Appendix.** Characteristics of Patients and Events at Sudden Death.

Case	Age (years)	Gender	Muscular impairment*	Pacemaker	Implantable defibrillator	Witnessed Collapse	Resuscitation Attempted	Initial Rhythm
1	39	Female	4	No	No	Yes	Yes	VT / VF‡
2	38	Female	3	No	No	No	No	-
3	62	Female	4	No	No	No	No	-
4	38	Female	4	No	No	No	No	-
5	48	Male	5	No	No	Yes	Yes§	VT / VF
6	33	Male	1,2	No	No	Yes	Yes	VT / VF
7	47	Female	4	No	No	Yes	Yes	Unknown
8	23	Female	1,2	No	No	Yes	Yes	Unknown
9	48	Female	5	No	No	No	No	-
10	54	Male	5	No	No	Yes	No¶	Bradycardia / 3 <sup>rd</sup> degree heart block
11	64	Male	5	Yes	No	Yes	No¶	VT / VF
12	77	Male	4	No	No	No	No	-
13	45	Female	4	Yes	No	Yes	Yes	Asystole (pacing spikes)
14	55	Male	4	Yes	No	Yes	Yes	VT / VF
15	58	Female	1,2	No	No	Yes	Yes	VT / VF

16	40	Female	5	No	No	No	No	-
17	45	Male	4	Yes	No	Yes	Yes	Asystole (pacing spikes)
18	76	Female	4	No	No	Yes	Yes	Asystole
19	51	Female	3	No	No	Yes	No¶¶	-
20	67	Female	3	No	No	Yes	Yes	Asystole
21	49	Female	5	No	No	Yes	No¶¶	VT / VF
22	47	Male	3	No	Yes	No	No	-
23	55	Male	3	Yes	No	Yes	Yes	VT / VF
24	54	Male	3	No	Yes	No	Yes	Asystole (pacing spikes)†
25	56	Male	3	No	No	No	Yes	Pulseless electrical activity
26	52	Male	3	No	No	No	Yes	Asystole
27	56	Male	4	No	No	Yes	Yes	VT / VF

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\*See text for definition of muscular impairment rating scale. Rating based on findings at last follow-up prior to sudden death.

‡VT / VF denotes a ventricular tachyarrhythmia observed on electrocardiogram monitor either ventricular tachycardia or fibrillation.

§Emergency response, rhythm evaluated, cardiopulmonary resuscitation initiated, and defibrillation with post-defibrillation asystole. Resuscitation then terminated per discussion with spouse based on patient's advance health care directive.

¶Emergency response and rhythm evaluated but no further resuscitation attempted per patients' advance health care directives.

¶¶No emergency response, rhythm evaluation, or resuscitation attempt due to location of witnessed collapse in a foreign country.

†Postmortem implantable cardioverter-defibrillator interrogation was evaluated. Antitachycardia pacing and cardioversion initially for atrial tachyarrhythmias and later defibrillation for ventricular tachyarrhythmias was observed over a period of one hour prior to death. The final event was defibrillation for a rapid ventricular tachyarrhythmia with subsequent asystole despite pacing spikes (rhythm observed by emergency responders).