

Supplemental Material

Table S1. Structural Evaluation of the Mitral, Tricuspid, Aortic, and Pulmonic Valves by Cohort.*

	Dystrophinopathies (n=23)	LGMD (n=21)	DM1 (n=50)	FSHD (n=12)
Mitral Valve	1 Mild Thickening, 2 Mild Prolapse	1 Mild Thickening	Normal	1 Mild Thickening
Tricuspid Valve	Normal	Normal	Normal	Normal
Aortic Valve	Normal	Normal	Normal	Normal
Pulmonic Valve	Normal	Normal	Normal	Normal

DM1 indicates type 1 myotonic dystrophy; FSHD, facioscapulohumeral muscular dystrophy; and LGMD, limb-girdle muscular dystrophy. *Data obtained from the 106 patients that received an echocardiogram study at baseline.

Table S2. Comparing 12-Lead Electrocardiogram and Systolic Function Data of Type 1 Myotonic Dystrophy Patients with Type 2 Myotonic Dystrophy Patients.*

	DM1 (n=60)	DM2 (n=7)	P Value
Heart Rate, bpm	68 (63.3-75)	69.5 (66-74.5)	0.61
PR Interval, ms	193 (178.3-223)	152 (144.5-173.5)	0.01
QRS Duration, ms	107 (96-118)	92 (86-95.5)	0.02
QT _c Interval, ms	412 (396-440)	399.5 (393-409.8)	0.37
1° AVB	18 (30)	0	
LAFB	11 (18.3)	0	
LBBB	6 (10)	0	
LVEF, %	55 (50-60)	60 (51.3-60)	0.52

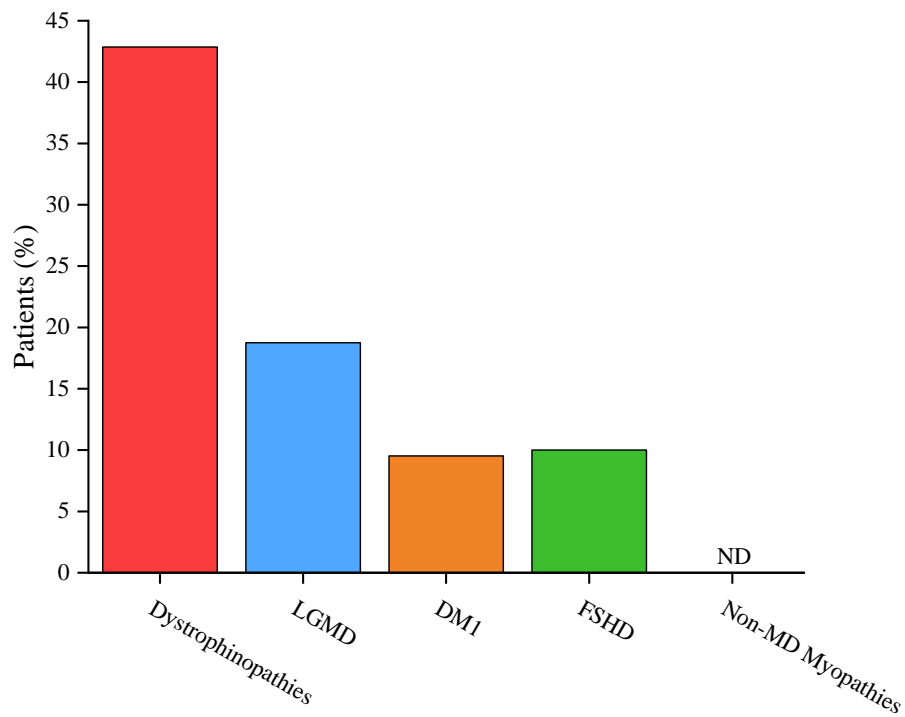
1° AVB indicates first-degree atrioventricular block; DM1, type 1 myotonic dystrophy; DM2, type 2 myotonic dystrophy; LAFB, left anterior fascicular block; LBBB, left bundle branch block; LVEF, left ventricular ejection fraction; PR Interval, duration of atrial depolarization; QRS Duration, duration of ventricular depolarization; and QT_c Interval, corrected duration between ventricular depolarization and repolarization. *12-lead electrocardiogram data was obtained from all 67 patients. Echocardiogram data was obtained from 54 patients (50 DM1 and four DM2 patients).

Table S3. Defined Maximum Tolerated Dose of Medications.*

Drug	Maximal Dose (mg)		Drug	Maximal Dose (mg)
ACEi			Beta-Blockers	
Enalapril	20		Bisoprolol	10
Lisinopril	40		Carvedilol	50
Perindopril	8		Metoprolol	200
Ramipril	10			
ARB			MRA	
Candesartan	32		Spironolactone	50
Irbesartan	300			
Telmisartan	80			
Valsartan	320			

ACEi indicates angiotensin converting enzyme inhibitor; ARB, angiotensin receptor blocker; and MRA, mineralocorticoid receptor antagonist. *Maximal dose defined as per the 2016 American Heart Association Guidelines.¹

Figure S1. Prevalence of myocardial fibrosis based on cardiac magnetic resonance imaging (MRI).



Patients screened by cardiac MRI that showed fibrosis as visualized by late gadolinium enhancement. DM1 indicates type 1 myotonic dystrophy; FSHD, facioscapulohumeral muscular dystrophy; LGMD, limb-girdle muscular dystrophy; and ND, not detected.

Supplemental Reference:

1. Yancy CW, Jessup M, Bozkurt B, Butler J, Casey DE, Colvin MM, Drazner MH, Filippatos G, Fonarow GC, Givertz MM, Hollenberg SM, Lindenfeld J, Masoudi FA, McBride PE, Peterson PN, Stevenson LW, Westlake C. 2016 ACC/AHA/HFSA Focused Update on New Pharmacological Therapy for Heart Failure: An Update of the 2013 ACCF/AHA Guideline for the Management of Heart Failure. *A Report of the American College of Cardiology/American Heart Association Task Force on Clinical Practice Guidelines and the Heart Failure Society of America.* 2016;68:1476-1488.