

Supplemental Material

Table. Online supplement presenting all candidate predictors assessed for disease progression. Mean±SD or Frequency (%) shown unless otherwise noted.

Variable*	n	Stable disease (n=99)	n	Progressive disease (n=28)	p [†]
Median age at diagnosis of cardiomyopathy, years (IQR)	99	1.1 (0.3-10.2)	28	7.5 (0.9-13.5)	0.04
Median time from diagnosis to baseline echo, years (IQR)	99	3.47 (0.56-6.84)	28	0.79 (0.27-2.97)	0.01
Median age at baseline echo, years (IQR)	99	8.83 (4.00-15.05)	28	9.39 (3.62-14.76)	0.74
Time between echos, years	90	0.81±0.31	17	0.73±0.30	0.31
Male	99	39 (39%)	28	17 (61%)	0.048
Race	99		28		0.06
White		69 (70%)		14 (50%)	
Other or unknown race		30 (30%)		14 (50%)	
Hispanic or Latino ethnicity	94	16 (17%)	27	2 (7%)	0.23
Symptom duration >0 months	98	15 (15%)	28	5 (18%)	0.74
Primary cause of dilated cardiomyopathy	99		28		0.57
Idiopathic		65 (66%)		20 (71%)	
Non-idiopathic		34 (34%)		8 (29%)	
Known or suspected familial cardiomyopathy	99	13 (13%)	28	6 (21%)	0.28

Variable*	n	Stable disease (n=99)	n	Progressive disease (n=28)	P [†]
Hospitalized in previous six months	99	26 (26%)	28	15 (54%)	0.008
No. of hospitalizations related to heart failure	99		28		0.006
0		86 (87%)		17 (61%)	
1		11 (11%)		6 (21%)	
2		2 (2%)		5 (18%)	
Failure to thrive in 6 months prior to enrollment	98	9 (9%)	28	6 (21%)	0.09
Feeding difficulties in 6 months prior to enrollment	99	11 (11%)	28	4 (14%)	0.53
Dyspnea in 6 months prior to enrollment	99	11 (11%)	27	12 (44%)	<0.001
Feeding difficulties or exercise intolerance in 6 months prior to enrollment	99	32 (32%)	28	18 (64%)	0.02
Edema in 6 months prior to enrollment	99	9 (9%)	28	3 (11%)	0.80
Syncope in 6 months prior to enrollment	98	5 (5%)	26	2 (8%)	0.61
Palpitations in 6 months prior to enrollment	91	9 (10%)	24	5 (21%)	0.15
Thrombo-embolic event in 6 months prior to enrollment	99	1 (1%)	28	1 (4%)	0.37

Variable*	n	Stable disease (n=99)	n	Progressive disease (n=28)	P [†]
Other symptoms in 6 months prior to enrollment	99	12 (12%)	28	1 (4%)	0.22
Intravenous inotropic medications in 6 months prior to enrollment	90	8 (9%)	27	7 (26%)	0.03
Ventricular tachyarrhythmia	9	6 (67%)	5	5 (100%)	0.96
Weight-for-age z-score	97	0.0±1.4	28	-0.2±1.8	0.46
Weight-for-age z-score	97		28		0.21
Z<-1		25 (26%)		11 (39%)	
-1≤Z≤1		53 (55%)		10 (36%)	
Z>1		19 (20%)		7 (25%)	
Median body surface area, m ² (IQR)	99	0.9 (0.7-1.7)	28	1.1 (0.6-1.6)	0.72
Body surface area-for-age z-score	98	0.1±1.3	28	0.2±1.9	0.77
End-diastolic short axis dimension, m-mode z-score	99	3.4±1.8	26	7.4±3.7	<0.001
End-diastolic short axis dimension, 2D z-score	99	3.6±2.4	27	7.8±4.3	<0.001
Posterior wall thickness to dimension ratio, m-mode z-score	98	-1.8±1.0	27	-2.6±1.0	<0.001

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Posterior wall thickness to dimension ratio, 2D z-score	99	-1.5±1.2	28	-2.2±1.3	0.007
LV mass z-score m-mode	99	1.3±1.5	26	3.2±2.2	<0.001
LV mass z-score, 2D	98	1.9±1.7	27	4.6±2.3	<0.001
Mass to volume ratio z-score	99	-1.0±1.1	28	-1.7±1.2	0.006
Cardiac index, 2D, L/min/m ²	99	4.0±1.2	28	4.1±1.4	0.56
Sphericity index z-score	99	1.0±1.3	28	2.0±1.4	0.001
Median eccentricity index (IQR)	99	0.93 (0.92-0.94)	28	0.92 (0.90-0.93)	0.003
Shortening fraction, m-mode %	99	21.7±6.4	27	12.8±4.8	<0.001
Median (IQR)	99	21.9 (18.0-27.0)	27	12.0 (8.7-16.7)	
Shortening fraction, m-mode z-score	99	-6.1±3.9	27	-12.1±4.8	<0.001
Shortening fraction, 2D %	99	18.7±6.2	28	11.6±5.3	<0.001
Median (IQR)	99	18.6 (14.3-23.7)	28	11.7 (8.3-13.2)	
Shortening fraction, 2D z-score	99	-5.1±1.9	28	-7.2±1.8	<0.001
Aortic ejection time, m-mode, msec	99	263.6±36.3	28	241.4±33.1	0.006
Heart rate / aortic ejection time, msec ^{1/2}	98	0.60±0.04	27	0.60±0.05	0.99

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Ejection fraction %	99	43.1±9.7	28	27.9±11.4	<0.001
Median (IQR)	99	44.0 (37.6-51.0)	28	27.1 (19.1-35.0)	
Ejection fraction z-score	99	-4.3±2.1	28	-7.5±2.4	<0.001
Mitral valve peak early velocity z-score*	69	0.3±1.1	16	-0.4±0.9	0.03
Mitral valve peak atrial velocity z-score*	69	0.3±1.2	16	0.0±1.4	0.31
Mitral valve early velocity/Atrial velocity z-score*	69	0.0±1.1	16	-0.1±1.2	0.83
Median mitral regurgitation jet width/BSA ^{0.5}	98	0.19 (0.00-0.29)	28	0.33 (0.21-0.46)	0.001
Severity of mitral valve regurgitation	98		28		0.004
None		33 (34%)		3 (11%)	
Mild		44 (45%)		9 (32%)	
Moderate		13 (13%)		8 (29%)	
Severe		8 (8%)		8 (29%)	
Severity of mitral valve regurgitation	98		28		<0.001
Non/Mild		77 (79%)		12 (43%)	
Moderate/Severe		21 (21%)		16 (57%)	
Tei Index	99	0.42±0.16	28	0.54±0.25	0.005

Variable*	n	Stable disease (n=99)	n	Progressive disease (n=28)	P [†]
Left lateral AV valve peak early diastolic velocity z-score*	86	-1.3±1.5	23	-2.6±1.7	<0.001
Left lateral AV valve peak atrial diastolic velocity z-score*	86	-0.6±2.0	23	-1.1±0.9	0.21
Left lateral AV valve peak systolic velocity z-score	99	-1.2±1.4	26	-2.3±1.2	<0.001
Septal AV valve peak early diastolic velocity z-score*	79	-1.9±1.2	21	-3.2±1.3	<0.001
Septal AV valve peak atrial diastolic velocity z-score*	79	-0.5±0.9	21	-1.0±1.1	0.03
Septal AV valve peak systolic velocity z-score	99	-1.4±2.1	27	-3.1±2.2	<0.001
Right lateral AV valve peak early diastolic velocity z-score*	67	-0.5±1.1	19	-1.2±0.9	0.02
Right lateral AV valve peak atrial diastolic velocity z-score*	67	-0.2±1.0	19	0.3±1.5	0.15
Right lateral AV valve peak systolic velocity right lateral AV valve z-score	98	-0.2±1.4	26	-1.1±1.4	0.002
Mitral valve average E/E' z-score*	67	2.2±3.4	15	6.5±7.0	0.004

Variable*	n	Stable disease (n=99)	n	Progressive disease (n=28)	P [†]
Left ventricular flow propagation velocity z-score	94	-0.3±0.9	28	-0.7±0.5	0.03
Mitral valve left lateral E/E' z-score*	69	2.2±3.3	15	7.4±8.0	0.002
Mitral valve septal E/E' z-score*	67	3.4±4.4	15	6.8±6.5	0.03

* Doppler variables which were only measurable in the absence of a summation wave were not considered for the stepwise multivariable model

All echocardiographic measures are from the left ventricle, where applicable.

†P value is from logistic regression comparing stable disease to significant disease progression.

AV=atrioventricular, IQR = interquartile range, echos=echocardiograms, LV=left ventricular