

Supplementary Online Content

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eTable 1. Baseline clinical characteristics of patients with 1 year or more of follow-up by BMI category with pathogenic/likely pathogenic variants

eTable 2. Clinical outcomes of patients with 1 year or more of follow-up by BMI category

eTable 3. Cox multivariable regression models for patients with pathogenic/likely pathogenic variants with 1 or more years of follow-up

eTable 4. Cox multivariable regression model for the overall composite end point for patients who performed a genetic test and had a single pathogenic/likely pathogenic mutation or were genotype negative with 1 year or more of follow-up

eFigure. Flow diagram summarizing patient selection in SHARE (March 2018 registry update)

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

eTable 1. Baseline clinical characteristics of patients with 1 year or more of follow-up by BMI category with pathogenic/likely pathogenic variants

	BMI Category P/LP				p
	Overall	<25	25-30	>30	
	1035	358 (34.6)	418 (40.4)	259 (25.0)	
Demographics					
US SHaRE Site, n (%)	440 (43)	103 (29)	170 (41)	167 (65)	<0.001
Age at Diagnosis	42±15	40±15	43±14	43±13	0.006
BMI	28±5	23±2	27±1	34±5	<0.001
Gender (men), n (%)	598 (58)	182 (51)	274 (66)	142 (55)	<0.001
Race					
White, n (%)	896 (87)	308 (86)	369 (88)	219 (85)	0.008
Black, n (%)	24 (2.3)	4 (1.1)	7 (1.7)	13 (5.0)	
Asian, n (%)	18 (1.7)	11 (3.1)	5 (1.2)	2 (0.8)	
Other, n (%)	97 (9.4)	35 (9.8)	37 (8.9)	25 (9.7)	
NYHA					
III/IV, n (%)	118 (11)	32 (9)	38 (9)	48 (19)	<0.001
Hypertension, n (%)	278 (27)	65 (18)	102 (24)	111 (43)	<0.001
Diabetes, n (%)	61 (5.8)	11 (3.1)	14 (3.3)	36 (13.8)	<0.001
SCD Family History, n (%)	266 (26)	87 (24)	103 (25)	76 (29)	0.325
Echocardiographic parameters					
LAD, mm	45±10	42±10	46±10	47±9	<0.001
MAX LVWT, mm	20±6	20±6	20±5	20±6	0.839
LVEF	65±9	64±10	65±9	66±9	0.026
LVOTO at Rest	12 [6-28]	12 [5-21]	10 [6-25]	16 [7-58]	0.001
LVOTO Valsalva	18 [8-61]	15 [8-48]	16 [7-54]	35 [10-100]	<0.001
Obstruction, n (%)	195 (19)	62 (17)	73 (18)	60 (23)	0.084

BMI: Body Mass Index; NYHA: New York Heart Association Functional Class; P/LP: Pathogenic/Likely Pathogenic; VUS: Variant of Unknown Significance; SCD: Sudden Cardiac Death; LAD: Left atrial diameter; LVWT: Left ventricle wall thickness; LVEF: Left ventricle ejection fraction, LVOTO: Left ventricle outflow tract obstruction.

eTable 2. Clinical outcomes of patients with 1 year or more of follow-up by BMI category

	BMI Category				P
	Overall	<25	25-30	>30	
	1035	358 (34.6)	418 (40.4)	259 (25.0)	
Demographics					
Age at last visit	51±15	50±16	51±15	53±13	0.014
Follow-Up	12±10	13±11	12±9	11±10	0.082
NYHA III/IV*, n (%)	116 (11)	35 (10)	41 (10)	40 (15)	0.038
SRT, n (%)	228 (22)	60 (17)	89 (21)	79 (31)	<0.001
NYHA III/IV*	39 (17)	8 (13)	14 (16)	17 (22)	0.197
ICD, n (%)	328 (32)	107 (30)	120 (29)	101 (39)	0.013
Appropriate Discharge	47 (14)	16 (15)	19 (16)	12 (12)	0.804
Clinical outcomes					
Atrial Fibrillation, n (%)	311 (30)	102 (28)	120 (29)	89 (34)	0.136
Syncope, n (%)	149 (14)	63 (18)	57 (14)	29 (11)	0.025
TIA/Stroke, n (%)	17 (1.7)	4 (1.1)	3 (0.7)	10 (3.8)	0.003
Death, n (%)	91 (8.8)	43 (12.0)	28 (6.7)	20 (7.7)	0.026
CVD, n (%)	54 (5.2)	24 (6.7)	18 (4.3)	12 (4.6)	0.243
Composite HF, n (%)	244 (24)	71 (20)	79 (19)	94 (36)	<0.001
Composite Overall, n (%)	477 (46)	148 (41)	182 (44)	147 (57)	<0.001
Composite Arrhythmia, n (%)	89 (9)	35 (9.8)	28 (6.7)	26 (10)	0.198
* = at last contact, BMI: Body Mass Index; NYHA: New York Heart Association Functional Class; P/LP: Pathogenic/Likely Pathogenic; VUS: Variant of Unknown Significance.					

eTable 3. Cox multivariable regression models for patients with pathogenic/likely pathogenic variants with 1 or more years of follow-up

	Composite Overall			HF-Composite			Atrial Fibrillation		
	HR	95% C.I.	p	HR	95% C.I.	p	HR	95% C.I.	p
BMI			0.004			<0.001			
25-30 vs <25	1.104	0.840-1.470		1.044	0.712-1.529				
>30 vs <25	1.557	1.180-2.091		1.784	1.237-2.571				
Age at Diagnosis	1.042	1.033-1.051	<0.001	1.037	1.025-1.049	<0.001	1.040	1.028-1.053	<0.001
Gender (men vs. women)				0.761	0.561-1.031	0.078			
LAD, mm	1.019	1.009-1.030	<0.001	1.023	1.011-1.037	<0.001	1.032	1.020-1.044	<0.001
Obstruction (yes vs. no)	1.812	1.429-2.297	<0.001	2.414	1.767-3.299	<0.001	1.509	1.076-2.115	0.017
BMI: Body Mass Index; LAD: Left Atrial Diameter.									

eTable 4. Cox multivariable regression model for the overall composite end point for patients who performed a genetic test and had a single pathogenic/likely pathogenic mutation or were genotype negative with 1 year or more of follow-up

	Composite Overall		
	HR	95% C.I.	p
BMI			0.001
25-30 vs <25	1.075	0.866-1.335	
>30 vs <25	1.659	1.324-2.078	
P/LP vs Genotype (-)	1.223	1.020-1.467	0.030
Age at Diagnosis	1.043	1.036-1.050	<0.001
LAD, mm	1.023	1.015-1.031	<0.001
Obstruction (yes vs. no)	1.612	1.348-1.929	<0.001

BMI: Body Mass Index; P/LP: Pathogenic/Likely Pathogenic gene variants; LAD: Left Atrial Diameter.

eFigure. Flow diagram summarizing patient selection in SHARE (March 2018 registry update)

