

Supplementary methods

Echocardiographic measurements were made according to current guidelines (1). Specifically, MLVWT was measured in the parasternal long-axis or parasternal short-axis views (2D or M-Mode) at end diastole. Extreme left ventricular hypertrophy (LVH) was defined as a MLVWT ≥ 30 mm or Z score ≥ 6 (1). LV diastolic dysfunction was assessed to be present if two out of four variables used to assess diastolic function (annular E' velocity, septal E' velocity, average E/E' ratio, LA diameter) were out of normal range for age and body surface area (2, 3). LV systolic dysfunction was defined as a LV fractional shortening $\leq 28\%$ or ejection fraction $\leq 55\%$ (4). Left ventricular outflow tract (LVOT) obstruction (LVOTO) was defined as a maximal LVOT gradient of ≥ 30 mmHg (1).

12-lead ECGs were analysed by one observer (T.R) unaware of the clinical details of the patients. ECGs were excluded if trace quality was poor. Age-specific normal values for ECG parameters were used (5). The following parameters were measured (average of 3 beats) from lead II, or V5 if quality of trace was poor: heart rate (bpm), QRS axis, PR interval (ms), Sokolow-Lyon score (SV1 or SV2 + RV5 or RV6 ≥ 35 mV)(6), QT interval (ms) and corrected QT interval (ms) using Bazett's formula(7) . The presence of the following parameters were described: dominant S wave in V4, pathological Q waves, pathological T wave inversion (>1 mm beyond V1 aged ≥ 14 years, or beyond V3 aged < 14 years), ST segment depression (≥ 2 mm in any lead), and ST segment elevation (≥ 2 mm in leads V1-V3, or ≥ 1 mm in all other leads).

Non-sustained ventricular tachycardia (NSVT) during ambulatory ECG monitoring was defined as three or more consecutive ventricular beats at a rate of greater than 120 beats/min with a duration of less than 30 seconds(1).

Supplementary tables

Supplementary table 1: Clinical characteristics of patients with and without heart failure symptoms

		Heart failure symptoms (n=11)	No heart failure symptoms (n=61)	P value
Baseline clinical assessment (n=72)	Any symptoms	11 (100%)	13 (21.3%)	<0.001
	Mean MLVWT (+/-SD)	12.88 (2.03)	12.77 (2.65)	0.9105
	Concentric hypertrophy	9 (81.8%)	56 (91.8%)	0.304
	LVOT obstruction (n=67)	0 (0%)	2 (3.5%)	0.572
	Systolic impairment (n=49)	2 (33.3%)	5 (9.3%)	0.093
	Diastolic impairment (n=39)	1 (25.0%)	5 (14.3%)	0.574
			Heart failure symptoms (n=11)	No heart failure symptoms (n=65)
Last clinical assessment (n=76)	Any symptoms	1 (12.5%)	10 (10.3%)	0.867
	Mean MLVWT (+/-SD)	12.82 (1.73)	13.01 (3.10)	0.883
	Concentric hypertrophy (n=64)	9 (90%)	59 (90.7%)	0.879
	LVOT obstruction (n=17)	1 (33.3%)	0 (0.0%)	0.026
	Systolic impairment (n=47)	1 (16.7%)	3 (7.3%)	0.443
	Diastolic impairment (n=33)	2 (50.0%)	8 (27.6%)	0.361

NYHA = New York Heart Association, LVOT = left ventricular outflow tract

Supplementary table 2: Baseline clinical characteristics of patients by era of presentation

	Pre-2000 (n=11)	2000 – 2009 (n=32)	2010 – 2018 (n=30)	P value
Age of HCM diagnosis	9.8 (+/-3.4)	10.6 (+/- 2.2)	11.4 (+/- 3.8)	0.013
Baseline cardiac assessment				
Any cardiac symptoms	3 (42.9%)	10 (33.3%)	10 (33.3%)	0.882
NYHA>2	2 (18.2%)	6 (18.8%)	2 (6.7%)	0.344
LVMWT (mm)	12.9 (+/- 2.2)	12.9 (+/- 2.9)	12.3 (+/- 2.8)	0.546
LVMWT Z score	7.3 (+/-2.3)	7.2 (+/-4.3)	6.2 (+/-3.1)	0.393
Systolic impairment	1 (16.7%)	4 (21.1%)	1 (4.2%)	0.23
Atrial arrhythmia	3 (27.3%)	5 (15.6%)	0 (0%)	0.025
Mortality	4 (36.4%)	4 (12.5%)	0 (0%)	0.004

Supplementary table 3: Clinical characteristics of patients with and without atrial arrhythmias

		Atrial arrhythmia (8)	No atrial arrhythmia (70)	P value
Baseline clinical assessment (n=78)	Any symptoms	2 (25%)	22 (31.4%)	0.153
	NYHA >2	1 (12.5%)	10 (14.3%)	0.891
	Mean MLVWT (+/-SD)	12.88 (2.03)	12.77 (2.65)	0.9105
	Concentric hypertrophy	7 (87.5%)	58 (82.9%)	0.581
	LVOT obstruction	0 (0%)	2 (2.9%)	0.877
	Systolic impairment	1 (12.5%)	5 (7.1%)	0.701
	Diastolic impairment	1 (12.5%)	5 (7.1%)	0.715
Last clinical assessment (n=76)	NYHA >2	1 (12.5%)	10 (10.3%)	0.867
	Mean MLVWT (+/-SD)	12.82 (1.73)	13.01 (3.10)	0.883
	Concentric hypertrophy	7 (87.5%)	51 (72.9%)	0.792
	LVOT obstruction	0 (0%)	1 (1.4%)	0.782
	Systolic impairment	3 (37.5%)	1 (1.43%)	<0.001
	Diastolic impairment	3 (37.5%)	7 (10.0%)	0.074

NYHA = New York Heart Association, LVOT = left ventricular outflow tract

References

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