

### 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  $\geq 30$ mm or Z score  $\geq 6$  (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  $\geq 30$ 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  $\geq 35$ 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  $\geq 14$  years, or beyond V3 aged  $< 14$  years), ST segment depression ( $\geq 2$ mm in any lead), and ST segment elevation ( $\geq 2$  mm in leads V1-V3, or  $\geq 1$ 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**

		<b>Heart failure symptoms (n=11)</b>	<b>No heart failure symptoms (n=61)</b>	<b>P value</b>
<b>Baseline clinical assessment (n=72)</b>	<b>Any symptoms</b>	11 (100%)	13 (21.3%)	<0.001
	<b>Mean MLVWT (+/-SD)</b>	12.88 (2.03)	12.77 (2.65)	0.9105
	<b>Concentric hypertrophy</b>	9 (81.8%)	56 (91.8%)	0.304
	<b>LVOT obstruction (n=67)</b>	0 (0%)	2 (3.5%)	0.572
	<b>Systolic impairment (n=49)</b>	2 (33.3%)	5 (9.3%)	0.093
	<b>Diastolic impairment (n=39)</b>	1 (25.0%)	5 (14.3%)	0.574
			<b>Heart failure symptoms (n=11)</b>	<b>No heart failure symptoms (n=65)</b>
<b>Last clinical assessment (n=76)</b>	<b>Any symptoms</b>	1 (12.5%)	10 (10.3%)	0.867
	<b>Mean MLVWT (+/-SD)</b>	12.82 (1.73)	13.01 (3.10)	0.883
	<b>Concentric hypertrophy (n=64)</b>	9 (90%)	59 (90.7%)	0.879
	<b>LVOT obstruction (n=17)</b>	1 (33.3%)	0 (0.0%)	0.026
	<b>Systolic impairment (n=47)</b>	1 (16.7%)	3 (7.3%)	0.443
	<b>Diastolic impairment (n=33)</b>	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
<b>Age of HCM diagnosis</b>	9.8 (+/-3.4)	10.6 (+/- 2.2)	11.4 (+/- 3.8)	0.013
<b>Baseline cardiac assessment</b>				
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
<b>Atrial arrhythmia</b>	3 (27.3%)	5 (15.6%)	0 (0%)	0.025
<b>Mortality</b>	4 (36.4%)	4 (12.5%)	0 (0%)	0.004

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

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

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

## References

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