

Title: Real-World Effectiveness of Mavacamten in Canada: Insights from the MAVEN Study

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Supplementary Table S1. Mavacamten effectiveness and safety in monotherapy subgroup

Demographics and Clinical Characteristics						
Measurement at Baseline	N	All Patients (N=115)	N	Mavacamten + HCM Background Therapy (N=93)	N	Mavacamten Monotherapy (N=22)
Age ¹ (years), median (range)	65	63.0 (22-87)	50	60.9 (22-87)	15	70.4 (53-82)
NYHA Class II, n (%)	115	66 (57.4%)	93	55 (59.1 %)	22	11 (50.0%)
vLVOT gradient (mmHg), median (IQR)	95	78.0 (60, 100)	76	77.5 (60, 100)	19	91.0 (56, 100)
rLVOT gradient (mmHg), median (IQR)	106	44.0 (16, 75)	86	47.0 (20, 77)	20	23.0 (9, 63)
LVEF (%), median (IQR)	111	65.0 (60, 70)	89	65.0 (60, 70)	22	64.0 (60, 70)
Effectiveness and Safety						
Measurement at Data Cut-off	N	All Patients (N=115)	N	Mavacamten + HCM Background Therapy (N=93)	N	Mavacamten Monotherapy (N=22)
Improvement in ≥ 1 NYHA class, n (%)	99	72 (72.7%)	81	59 (72.8%)	18	13 (72.2%)
Change in vLVOT gradient from baseline (mmHg), median (IQR)	93	-51.0 (-83, -24)	74	-49.5 (-78, -24)	19	-64.0 (-89, -24)
vLVOT ≤ 30 mmHg, n (%)	112	75 (67.0%)	91	58 (63.7%)	21	17 (81.0%)
Change in rLVOT gradient from baseline (mmHg), median (IQR)	98	-25.5 (-61, -8)	79	-31.0 (-66, -9)	19	-8.0 (-51, 0)
rLVOT ≤ 30 mmHg, n (%)	106	95 (89.6 %)	85	75 (88.2 %)	21	20 (95.2 %)
Mavacamten discontinuation due to TEAEs, n (%)	115	3 (2.6%)	93	3 (3.2%)	22	0 (0%)
Change in LVEF from baseline, median (IQR)	109	-3.0 (-8, 0)	88	-4.0 (-8, 0)	21	-1.0 (-8, 3)

¹Actual age data was missing for 50 patients.

HCM: hypertrophic cardiomyopathy; IQR: interquartile range; LVEF: left ventricular ejection fraction; NYHA: New York Heart Association; rLVOT: Resting Left Ventricular Outflow Tract; TEAEs: treatment-emergent adverse events; vLVOT: left ventricular outflow tract gradient with Valsalva manoeuvre.

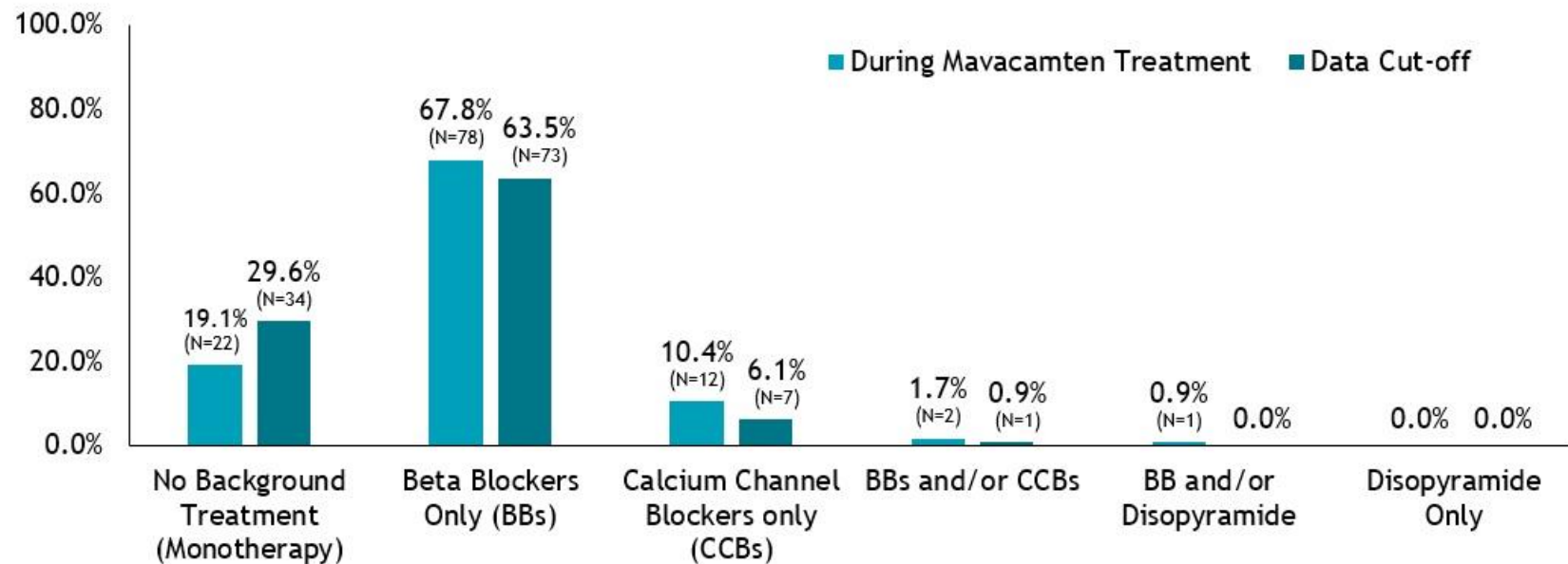
Supplementary Table S2. Mavacamten effectiveness and safety in subgroup of patients with LVOT < 20 mmHg during treatment initiation

Demographics and Clinical Characteristics						
Measurement at Baseline	N	All Patients (N=115)	N	Patient with LVOT < 20 mmHg No Dose Change (N=24)	N	Patient with LVOT < 20 mmHg Dose Change (N=28)
Age ¹ (years), median (range)	65	63.0 (22-87)	24	68.6 (58-72)	28	62.4 (54-69)
NYHA Class II, n (%)	115	66 (57.4%)	24	14 (58.3%)	28	20 (71.4%)
vLVOT gradient (mmHg), median (IQR)	95	78.0 (60, 100)	20	75.0 (55, 93)	24	91.5 (46, 133)
rLVOT gradient (mmHg), median (IQR)	106	44.0 (16, 75)	24	40.0 (20, 61)	26	39.0 (10, 72)
LVEF (%), median (IQR)	111	65.0 (60, 70)	23	65.0 (60, 70)	26	64.0 (60, 70)
Effectiveness and Safety						
Measurement at Data Cut-off	N	All Patients (N=115)	N	No Dose Change (N=24)	N	Dose Change (N=28)
Improvement in ≥ 1 NYHA class, n (%)	99	72 (72.7%)	22	22 (100%)	26	17 (65.4%)
Change in vLVOT gradient from baseline (mmHg), median (IQR)	93	-51.0 (-83, -24)	20	-50.0 (-79, -32)	24	-69.5 (-114, -21)
vLVOT ≤ 30 mmHg, n (%)	112	75 (67.0%)	24	18 (75%)	28	25 (89.3%)
Change in rLVOT gradient from baseline (mmHg), median (IQR)	98	-25.5 (-61, -8)	23	-35.0 (-66, -8)	26	-30.5 (-70, 0)
rLVOT ≤ 30 mmHg, n (%)	106	95 (89.6 %)	23	14 (58.3 %)	28	12 (46.2 %)
Mavacamten discontinuation due to TEAEs, n (%)	115	3 (2.6%)	24	0 (0%)	28	2 (7.1%)
Change in LVEF from baseline, median (IQR)	109	-3.0 (-8, 0)	23	-3.0 (-7, 0)	26	-5.0, (-10, 0)

¹Actual age data was missing for 50 patients.

HCM: hypertrophic cardiomyopathy; IQR: interquartile range; LVEF: left ventricular ejection fraction; NYHA: New York Heart Association; TEAEs: treatment-emergent adverse events; vLVOT: left ventricular outflow tract gradient with Valsalva manoeuvre.

Supplementary Figure S1. Background Hypertrophic cardiomyopathy (HCM) Medication During Mavacamten Treatment and at Data Cut-Off

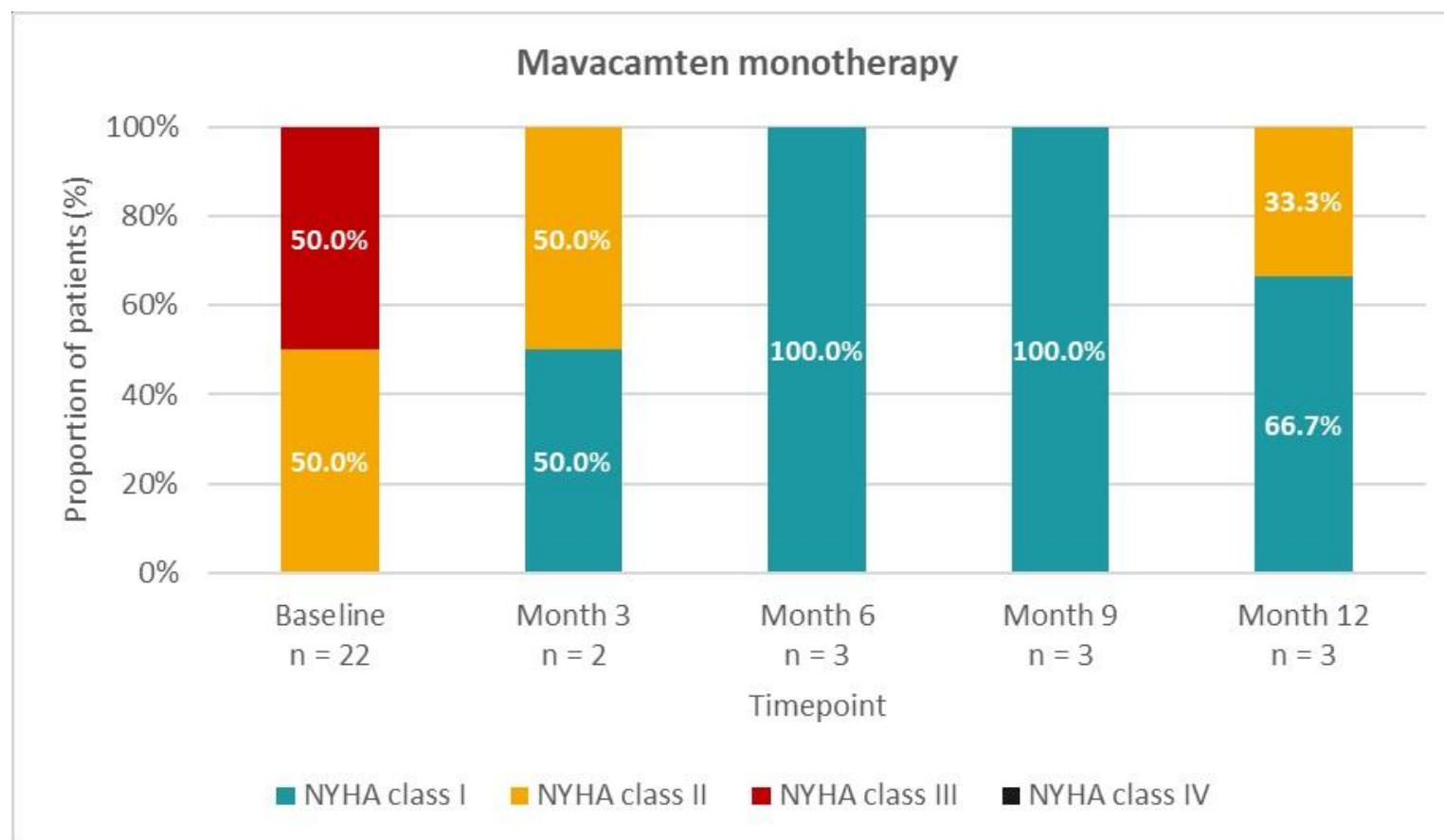


Supplementary Figure S1. Background HCM Medication During Mavacamten Treatment and at Data Cut-off:

Shows the proportion of patients receiving mavacamten as monotherapy, or concomitantly with BBs only, with CCBs only, with both BBs and/or CCBs, or with BBs and/or disopyramide, received either concurrently or sequentially.

“**During mavacamten treatment**” includes HCM medications used concomitantly with mavacamten, including medications discontinued on the same day as or after mavacamten initiation. “**Data cut-off**” represents each patient’s latest available assessment. Note: Only concomitant HCM medication categories were collected; specific medication names and dosages were not captured.

Supplementary Figure S2. New York Heart Association (NYHA) Functional Class at Selected Timepoints in Monotherapy Subgroup Cohort



Supplementary Figure S2. NYHA Functional Class at Selected Timepoints in Monotherapy Subgroup Cohort:

Shows the distribution of NYHA functional class among patients receiving mavacamten monotherapy at the selected assessment timepoints. At baseline (n=22), 50.0% of patients were NYHA Class II and 50.0% were NYHA Class III. NYHA Class I was observed at subsequent selected assessed timepoints; however, the number of patients with available follow-up assessments was limited (n=2-3).