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SUPPLEMENTARY MATERIAL

Magnetic resonance imaging evaluation of the effects of myosin inhibitors (mavacamten and aficamten) in hypertrophic cardiomyopathy: a systematic review and case report

Carolina Bernardes,¹ João Gomes Carvalho,^{1,2} Patrícia Rodrigues,^{1,3} Manuela França^{1,2}

¹School of Medicine and Biomedical Sciences, University of Porto; ²Radiology Department, Santo António University Hospital Centre of Porto; ³Cardiology Department, Santo António University Hospital Centre of Porto, Portugal

Correspondence: Carolina Bernardes, School of Medicine and Biomedical Sciences, University of Porto (ICBAS-UP), Rua Jorge Viterbo Ferreira 228, 4050-513 Porto, Portugal.

Tel.: +351 933423820. E-mail: carolinabernardes010@gmail.com

Key words: mavacamten, aficamten, myosin inhibitors, cardiac magnetic resonance, hypertrophic cardiomyopathy.

Supplementary Table 1. Research question.

Research Question (PICO)

Population: Adult patients with hypertrophic cardiomyopathy.
Intervention: Treatment with cardiac myosin inhibitors, either aficamten or mavacamten.
Comparison: Routine treatment.
Outcome: Changes in cardiac magnetic resonance imaging.

Supplementary Table 2. Inclusion and exclusion criteria.

Population

Inclusion criteria: Adult patients (> 18 years) diagnosed with hypertrophic cardiomyopathy.
Exclusion criteria: Not meeting inclusion criteria.

Intervention

Inclusion criteria: Treatment with aficamten or mavacamten.
Exclusion criteria: Not meeting inclusion criteria.

Comparison

Inclusion criteria: Routine treatment or any alternative control approach.
Exclusion criteria: Not applicable.

Outcome

Inclusion criteria: magnetic resonance imaging characteristics/parameters assessment during treatment with CMIs.
Exclusion criteria: Not meeting inclusion criteria.

Study type

Inclusion criteria: randomized controlled trials (RCTs).
Exclusion criteria: Not meeting inclusion criteria.

Date

Inclusion criteria: No restrictions.
Exclusion criteria: No restrictions.

Location

Inclusion criteria: No restrictions
Exclusion criteria: No restrictions.

Language

Inclusion criteria: English.
Exclusion criteria: None.

Supplementary Table 3. Search terms.

	Text Words	Controlled Vocabulary MeSH terms
Population	hypertrophic cardiomyopathy HCM hypertrophic obstructive cardiomyopathy obstructive hypertrophic cardiomyopathy oHCM	Cardiomyopathy, Hypertrophic
Intervention	MYK-461 CK-3773274 myosin inhibitor myosin inhibitors cardiac myosin inhibitor cardiac myosin inhibitors CMI CMIs aficamten mavacamten	
Outcome	magnetic resonance magnetic resonance imaging cardiac magnetic resonance cardiovascular magnetic resonance CMR	Magnetic Resonance Imaging

Supplementary Table 4. Search strategy.

Pubmed - 16/12/2024, 4 results:

("hypertrophic cardiomyopathy"[Text Word] OR "HCM"[Text Word] OR "hypertrophic obstructive cardiomyopathy"[Text Word] OR "obstructive hypertrophic cardiomyopathy"[Text Word] OR "oHCM"[Text Word] OR "cardiomyopathy, hypertrophic"[MeSH Terms]) AND ("MYK-461"[Text Word] OR "CK-3773274"[Text Word] OR "myosin inhibitor*" [Text Word] OR "cardiac myosin inhibitor*" [Text Word] OR "CMI"[Text Word] OR "CMIs"[Text Word] OR "aficamten"[Text Word] OR "mavacamten"[Text Word]) AND ("magnetic resonance"[Text Word] OR "Magnetic Resonance Imaging"[Text Word] OR "cardiac magnetic resonance"[Text Word] OR "cardiovascular magnetic resonance"[Text Word] OR "CMR"[Text Word] OR "Magnetic Resonance Imaging"[MeSH Terms]) AND (("randomized controlled trial"[Publication Type] OR "controlled clinical trial"[Publication Type] OR "randomized"[Title/Abstract] OR "placebo"[Title/Abstract] OR "drug therapy"[MeSH Subheading] OR "randomly"[Title/Abstract] OR "trial"[Title/Abstract] OR "groups"[Title/Abstract]) NOT ("animals"[MeSH Terms] NOT "humans"[MeSH Terms]))

Scopus - 16/12/2024, 32 results:

((TITLE-ABS-KEY ("hypertrophic cardiomyopathy" OR (hcm) OR "hypertrophic obstructive cardiomyopathy" OR "obstructive hypertrophic cardiomyopathy" OR (ohcm)) AND (TITLE-ABS-KEY ((myk-461) OR (ck-3773274) OR "myosin inhibitor*" OR "cardiac myosin inhibitor*" OR (cmi) OR (cmis) OR (aficamten) OR (mavacamten))) AND (TITLE-ABS-KEY ("magnetic resonance" OR "magnetic resonance imaging" OR "cardiac magnetic resonance" OR "cardiovascular magnetic resonance" OR (cmr)))) AND ((TITLE-ABS-KEY ("randomized controlled trial" OR "controlled clinical trial" OR (randomized) OR (placebo) OR "drug therapy" OR (randomly) OR (trial) OR (groups))) AND NOT (TITLE-ABS-KEY ((animals) AND NOT (humans))))

Cochrane Library - 16/12/2024, 7 results:

((("hypertrophic cardiomyopathy"):ti,ab,kw OR (HCM):ti,ab,kw OR ("hypertrophic obstructive cardiomyopathy"):ti,ab,kw OR ("obstructive hypertrophic cardiomyopathy"):ti,ab,kw OR (oHCM):ti,ab,kw OR [mh "Cardiomyopathy, Hypertrophic"]) AND ((MYK-461):ti,ab,kw OR (CK-3773274):ti,ab,kw OR ("myosin inhibitor"):ti,ab,kw OR ("myosin inhibitors"):ti,ab,kw OR ("cardiac myosin inhibitor"):ti,ab,kw OR ("cardiac myosin inhibitors"):ti,ab,kw OR (CMI):ti,ab,kw OR (CMIs):ti,ab,kw OR (aficamten):ti,ab,kw OR (mavacamten):ti,ab,kw) AND (("magnetic resonance"):ti,ab,kw OR ("magnetic resonance imaging"):ti,ab,kw OR ("cardiac magnetic resonance"):ti,ab,kw OR ("cardiovascular magnetic resonance"):ti,ab,kw OR (CMR):ti,ab,kw OR [mh "Magnetic Resonance Imaging"])

Web of Science - 16/12/2024, 13 results:

(ALL=("hypertrophic cardiomyopathy" OR HCM OR "hypertrophic obstructive cardiomyopathy" OR "obstructive hypertrophic cardiomyopathy" OR oHCM)) AND (ALL=(MYK-461 OR CK-3773274 OR "myosin inhibitor*" OR "cardiac myosin inhibitor*" OR CMI OR CMIs OR aficamten OR mavacamten)) AND (ALL=("magnetic resonance" OR "magnetic resonance imaging" OR "cardiac magnetic resonance" OR "cardiovascular magnetic resonance" OR CMR)) AND (ALL=(("randomized controlled trial" OR "controlled clinical trial" OR randomized OR placebo OR "drug therapy" OR randomly OR trial OR groups) NOT (animals NOT humans)))

ScienceDirect - 16/12/2024, 183 results:

("hypertrophic" AND "cardiomyopathy") AND (aficamten OR mavacamten) AND ("magnetic resonance" OR CMR) AND (randomized OR controlled OR trial)

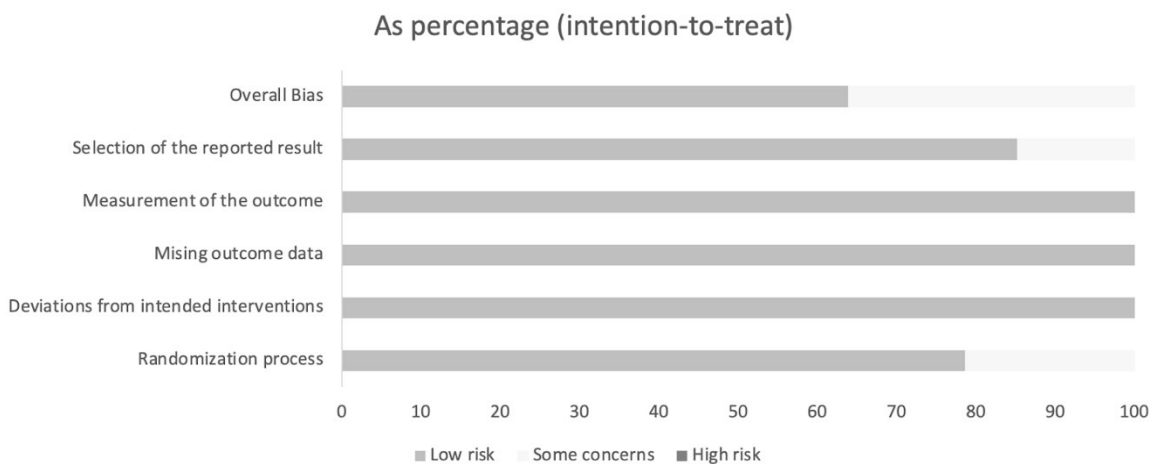
Supplementary Table 5. Risk of bias assessment.

Intention-to-treat	StudyID	Outcome	D1	D2	D3	D4	D5	Overall
	SEQUOIA-HCM CMR Substudy	Change from baseline in LV mass index	+	+	+	+	+	+
	SEQUOIA-HCM CMR Substudy	Change from baseline in LV maximal wall thickness	+	+	+	+	+	+
	SEQUOIA-HCM CMR Substudy	Change from baseline in maximum LA volume index	+	+	+	+	+	+
	SEQUOIA-HCM CMR Substudy	Change from baseline in LV end-diastolic volume index	+	+	+	+	+	+
	SEQUOIA-HCM CMR Substudy	Change from baseline in LV end-systolic volume index	+	+	+	+	+	+
	SEQUOIA-HCM CMR Substudy	Change from baseline in mitral regurgitation volume	+	+	+	+	+	+
	SEQUOIA-HCM CMR Substudy	Change from baseline in mitral regurgitation fraction	+	+	+	+	+	+
	SEQUOIA-HCM CMR Substudy	Change from baseline in LGE mass	+	+	+	+	+	+
	SEQUOIA-HCM CMR Substudy	Change from baseline in LGE, % of LV mass	+	+	+	+	+	+
	SEQUOIA-HCM CMR Substudy	Change from baseline in LV extracellular volume fraction	+	+	+	+	+	+
	SEQUOIA-HCM CMR Substudy	Change from baseline in LV native T1	+	+	+	!	!	!
	SEQUOIA-HCM CMR Substudy	Change from baseline in LV mass	+	+	+	+	!	!
	SEQUOIA-HCM CMR Substudy	Change from baseline in LV ejection fraction	+	+	+	+	+	+
	SEQUOIA-HCM CMR Substudy	Change from baseline in LV extracellular volume mass index	+	+	+	+	!	!
	SEQUOIA-HCM CMR Substudy	Change from baseline in absolute myocyte mass index	+	+	+	+	!	!
	EXPLORER-CN	Change from baseline in LV mass index	!	+	+	+	+	!
	EXPLORER-CN	Change from baseline in LV mass	!	+	+	+	+	!
	EXPLORER-CN	Change from baseline in LV maximal wall thickness	!	+	+	+	+	!
	EXPLORER-CN	Change from baseline in maximum LA volume index	!	+	+	+	+	!
	EXPLORER-CN	Change from baseline in minimum LA volume index	!	+	+	+	+	!
	EXPLORER-HCM CMR substudy	Change from baseline in LV mass index	+	+	+	+	+	+
	EXPLORER-HCM CMR substudy	Change from baseline in LV mass	+	+	+	+	+	+
	EXPLORER-HCM CMR substudy	Change from baseline in absolute intracellular myocardial mass index	+	+	+	+	+	+
	EXPLORER-HCM CMR substudy	Change from baseline in LV maximal wall thickness	+	+	+	+	+	+
	EXPLORER-HCM CMR substudy	Change from baseline in LV ejection fraction	+	+	+	+	+	+
	EXPLORER-HCM CMR substudy	Change from baseline in myocardial contractile fraction	+	+	+	+	+	+
	EXPLORER-HCM CMR substudy	Change from baseline in maximum LA volume index	+	+	+	+	+	+
	EXPLORER-HCM CMR substudy	Change from baseline in LGE	+	+	+	+	+	+
	EXPLORER-HCM CMR substudy	Change from baseline in extracellular volume fraction	+	+	+	+	+	+

+ Low risk
! Some concerns
- High risk

D1 Randomisation process
 D2 Deviations from the intended interventions
 D3 Missing outcome data
 D4 Measurement of the outcome
 D5 Selection of the reported result

CMR, cardiac magnetic resonance; LV, left ventricular; LGE, late gadolinium enhancement; N/A, not available; LA, left atrial.



Supplementary Figure 1. Risk of bias assessment.