DGK CARDIO UPDATE 2024

16. und 17. Februar
23. und 24. Februar

Kardiomyopathien, Speicherekrankungen, Myokarditis

DGK CARDIO UPDATE 2024

16. und 17. Februar
23. und 24. Februar

Keine Interessenskonflikte

ESC
European Society of Cardiology
European Heart Journal (2023) 44, 3503–3626
https://doi.org/10.1093/eurheartj/ehad194

ESC GUIDELINES

2023 ESC Guidelines for the management of cardiomyopathies

Developed by the task force on the management of cardiomyopathies of the European Society of Cardiology (ESC)

Authors/Task Force Members: Elena Arbelo ✉*†, (Chairperson) (Spain), Alexandros Protonotarios ‡, (Task Force Co-ordinator) (United Kingdom), Juan R. Gimeno ‡, (Task Force Co-ordinator) (Spain), Eloisa Arbustini (Italy), Connie R. Bezzina (Netherlands), Roberto Barriales-Villa (Spain), Cristina Basso (Italy), Nico A. Blom[1] (Netherlands), Elena Biagini (Italy), Rudolf A. de Boer (Netherlands), Tim De Winter (Belgium), Perry M. Elliott (United Kingdom), Marcus Flather (United Kingdom), Pablo Garcia-Pavia (Spain), Kristina H. Haugaa (Sweden), Jodie Ingles (Australia), Ruxandra Oana Jurcut (Romania), Sabine Klaassen (Germany), Giuseppe Limongelli (Italy), Bart Loeys[2] (Belgium), Jens Mogensen (Denmark), Iacopo Olivotto (Italy), Antonis Pantazis (United Kingdom), Sanjay Sharma (United Kingdom), J. Peter Van Tintelen (Netherlands), James S. Ware (United Kingdom), Juan Pablo Kaski ✉*†, (Chairperson) (United Kingdom), and ESC Scientific Document Group

✉ Corresponding authors: Elena Arbelo, Arrhythmia Section, Cardiology Department, Hospital Clínic, Universitat de Barcelona, Barcelona, Spain, IDIBAPS, Institut d'Investigació August Pi i Sunyer (IDIBAPS), Barcelona, Spain, Centro de Investigación Biomédica en Red de Enfermedades Cardiovasculares (CIBERCV), Madrid, Spain, and European Reference Network for Rare, Low Prevalence and Complex Diseases of the Heart, ERN GUARD-Heart, Barcelona, Spain. Tel: +34 93 22 75 55 11, E-mail: elenaarbelo@secardiologia.es; and Juan Pablo Kaski, Centre for Paediatric Inherited and Rare Cardiovascular Disease, University College London, Institute of Cardiovascular Science, London, United Kingdom and Centre for Inherited Cardiovascular Diseases, Great Ormond Street Hospital, London, United Kingdom. Tel: +44 78 29 68 39, E-mail: j.kaski@ucl.ac.uk

† The two Chairpersons contributed equally to the document and are joint corresponding authors.

‡ The two Task Force Co-ordinators contributed equally to the document.

Author/Task Force Member affiliations are listed in author information.

[1] Representing the Association for European Paediatric and Congenital Cardiology (AEPC)

[2] Representing the European Society of Human Genetics (ESHG)

ESC Clinical Practice Guidelines (CPG) Committee: listed in the Appendix.

ESC subspecialty communities having participated in the development of this document:

Associations: Association of Cardiovascular Nursing & Allied Professions (ACNAP), European Association of Cardiovascular Imaging (EACVI), European Association of Preventive Cardiology (EAPC), European Heart Rhythm Association (EHRA), Heart Failure Association (HFA).

Councils: Council on Cardiovascular Genomics.

Working Groups: Development Anatomy and Pathology, Myocardial and Pericardial Diseases.

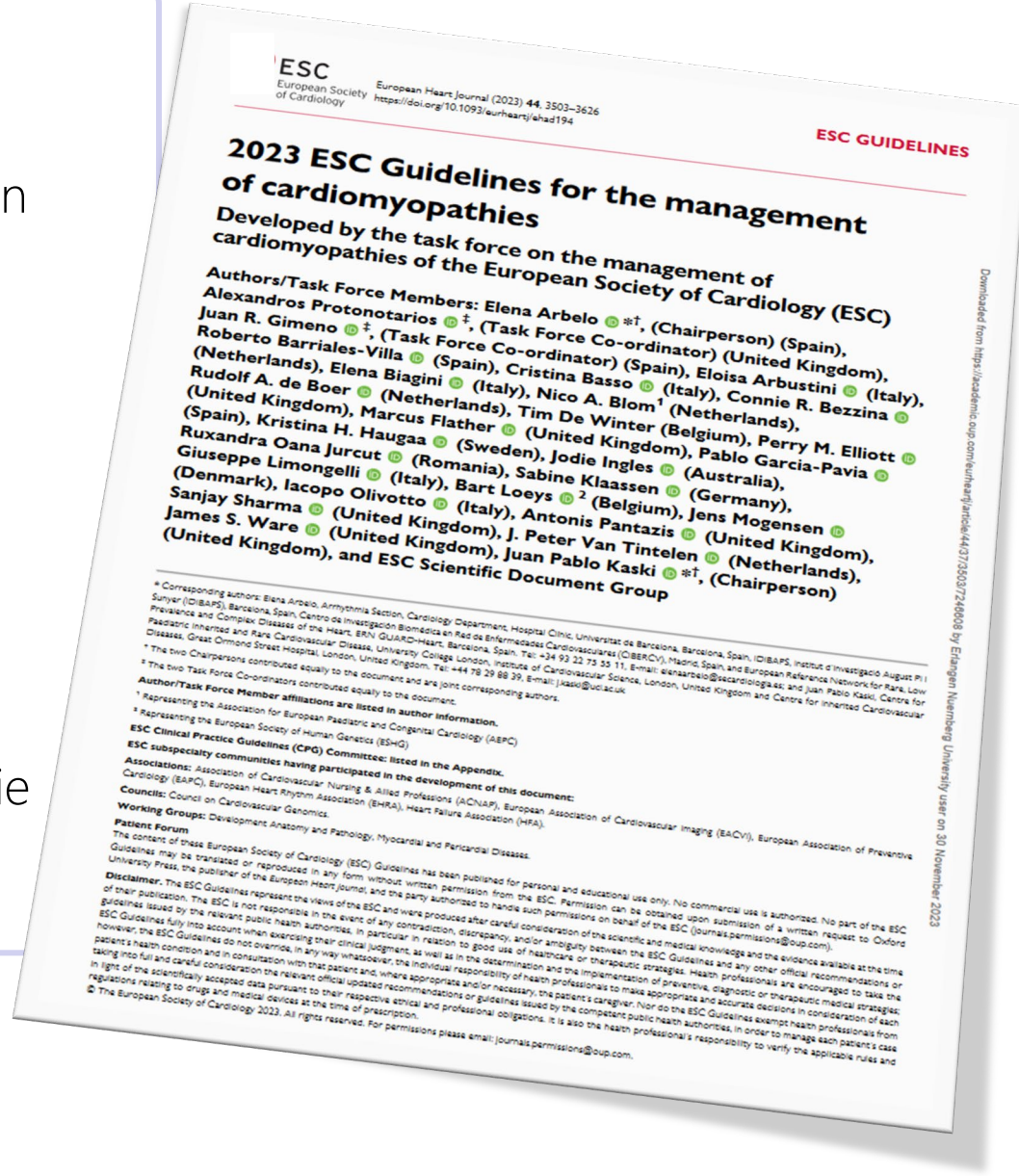
Patient Forum

Definition

Myokardiale Erkrankung mit einem strukturell und funktionell abnormen Herzmuskel, ohne Vorliegen von

- KHK
- Hypertonie
- Klappenfehlern
- angeborenen Herzfehlern

von ausreichendem Ausmaß, um die myokardiale Störung zu erklären



ESC European Society of Cardiology

European Heart Journal (2023) 44, 3503–3626
https://doi.org/10.1093/eurheartj/ehad194

ESC GUIDELINES

2023 ESC Guidelines for the management of cardiomyopathies

Developed by the task force on the management of cardiomyopathies of the European Society of Cardiology (ESC)

Authors/Task Force Members: Elena Arbelo *†, (Chairperson) (Spain), Alexandros Protonotarios ‡, (Task Force Co-ordinator) (United Kingdom), Juan R. Gimeno ‡, (Task Force Co-ordinator) (Spain), Eloisa Arbustini (Italy), Roberto Barriales-Villa (Spain), Cristina Basso (Italy), Connie R. Bezzina (Netherlands), Elena Biagini (Italy), Nico A. Blom[1] (Netherlands), Rudolf A. de Boer (Netherlands), Tim De Winter (Belgium), Perry M. Elliott (United Kingdom), Marcus Flather (United Kingdom), Pablo Garcia-Pavia (Spain), Kristina H. Haugaa (Sweden), Jodie Ingles (Australia), Ruxandra Oana Jurcut (Romania), Sabine Klaassen (Germany), Giuseppe Limongelli (Italy), Bart Loeys[2] (Belgium), Jens Mogensen (Denmark), Iacopo Olivotto (Italy), Antonis Pantazis (United Kingdom), Sanjay Sharma (United Kingdom), J. Peter Van Tintelen (Netherlands), James S. Ware (United Kingdom), Juan Pablo Kaski *†, (Chairperson) (United Kingdom), and ESC Scientific Document Group

* Corresponding authors: Elena Arbelo, Arrhythmia Section, Cardiology Department, Hospital Clínic, Universitat de Barcelona, Barcelona, Spain, IDIBAPS, Institut d'Investigació August Pi i Sunyer (IDIBAPS), Barcelona, Spain, Centro de Investigación Biomédica en Red de Enfermedades Cardiovasculares (CIBERCV), Madrid, Spain, and European Reference Network for Rare, Low Prevalence and Complex Diseases of the Heart, ERN GUARD-Heart, Barcelona, Spain. Tel: +34 93 22 75 55 11, E-mail: elenaarbelo@secardiologia.es; and Juan Pablo Kaski, Centre for Paediatric Inherited and Rare Cardiovascular Diseases, University College London, Institute of Cardiovascular Science, London, United Kingdom and Centre for Inherited Cardiovascular Diseases, Great Ormond Street Hospital, London, United Kingdom. Tel: +44 78 29 88 39, E-mail: j.kaski@ucl.ac.uk

† The two Chairpersons contributed equally to the documents and are joint corresponding authors.

‡ The two Task Force Co-ordinators contributed equally to the document.

Author/Task Force Member affiliations are listed in author information.

[1] Representing the Association for European Paediatric and Congenital Cardiology (AEPC)

[2] Representing the European Society of Human Genetics (ESHG)

ESC Clinical Practice Guidelines (CPG) Committee: listed in the Appendix.

ESC subspecialty communities having participated in the development of this document:

Associations: Association of Cardiovascular Nursing & Allied Professions (ACNAP), European Association of Cardiovascular Imaging (EACVI), European Association of Preventive Cardiology (EAPC), European Heart Rhythm Association (EHRA), Heart Failure Association (HFA).

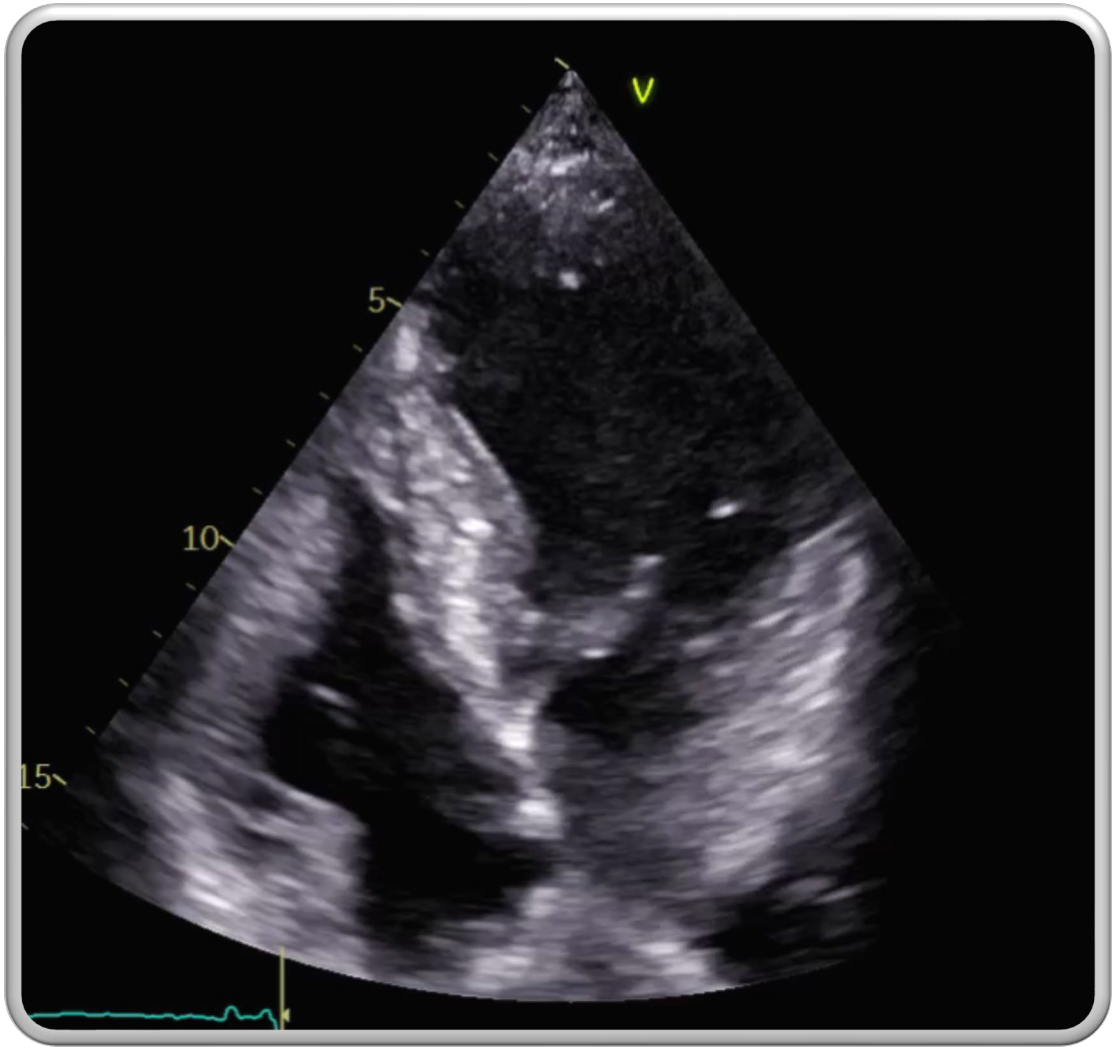
Councils: Council on Cardiovascular Genomics.

Working Groups: Development Anatomy and Pathology, Myocardial and Pericardial Diseases.

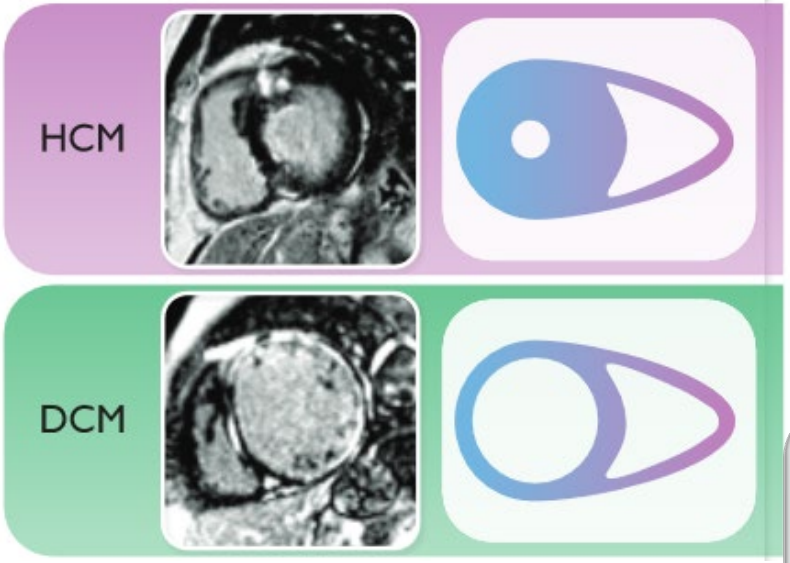
Patient Forum

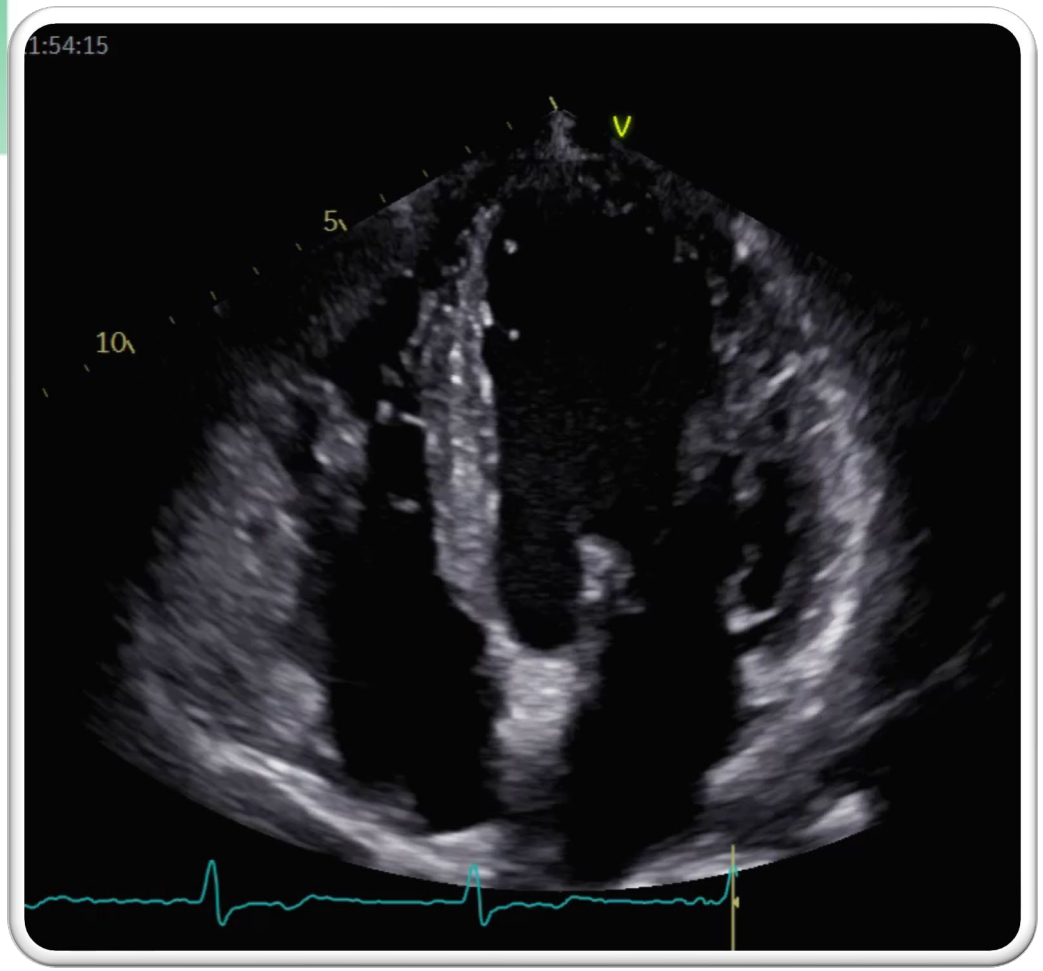
Disclaimer. The ESC Guidelines represent the views of the ESC and were produced after careful consideration of the scientific and medical knowledge and the evidence available at the time of their publication. The ESC is not responsible in the event of any contradiction, discrepancy, and/or ambiguity between the ESC Guidelines and any other official recommendations or guidelines issued by the relevant public health authorities. In particular in relation to good use of healthcare or therapeutic strategies. Health professionals are encouraged to take the ESC Guidelines fully into account when exercising their clinical judgment, as well as in the determination and the implementation of preventive, diagnostic or therapeutic medical strategies; however, the ESC Guidelines do not override, in any way whatsoever, the individual responsibility of health professionals to make appropriate and accurate decisions in consideration of each patient's health condition and in consultation with that patient and, where appropriate and/or necessary, the patient's caregiver. Nor do the ESC Guidelines exempt health professionals from taking into full and careful consideration the relevant official updated recommendations or guidelines issued by the competent public health authorities, in order to manage each patient's case in light of the scientifically accepted data pursuant to their respective ethical and professional obligations. It is also the health professional's responsibility to verify the applicable rules and regulations relating to drugs and medical devices at the time of prescription.

Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

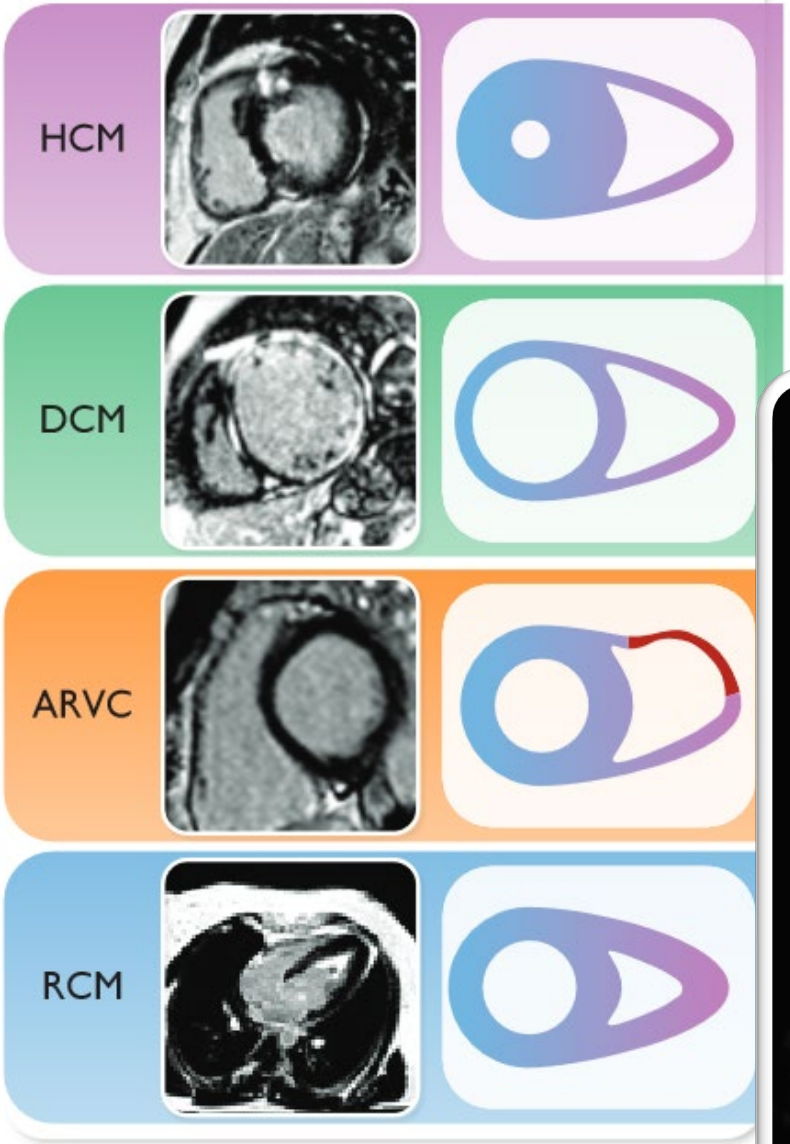


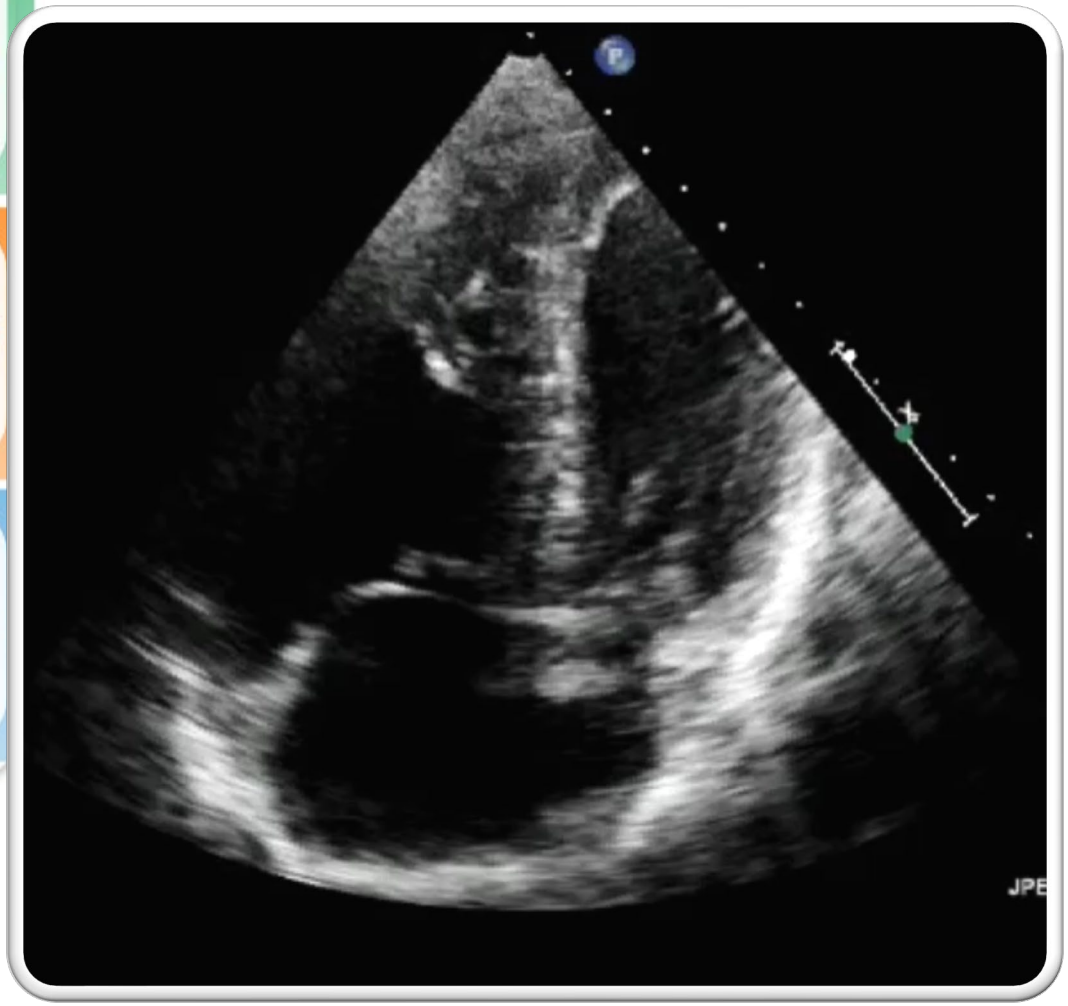
Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)



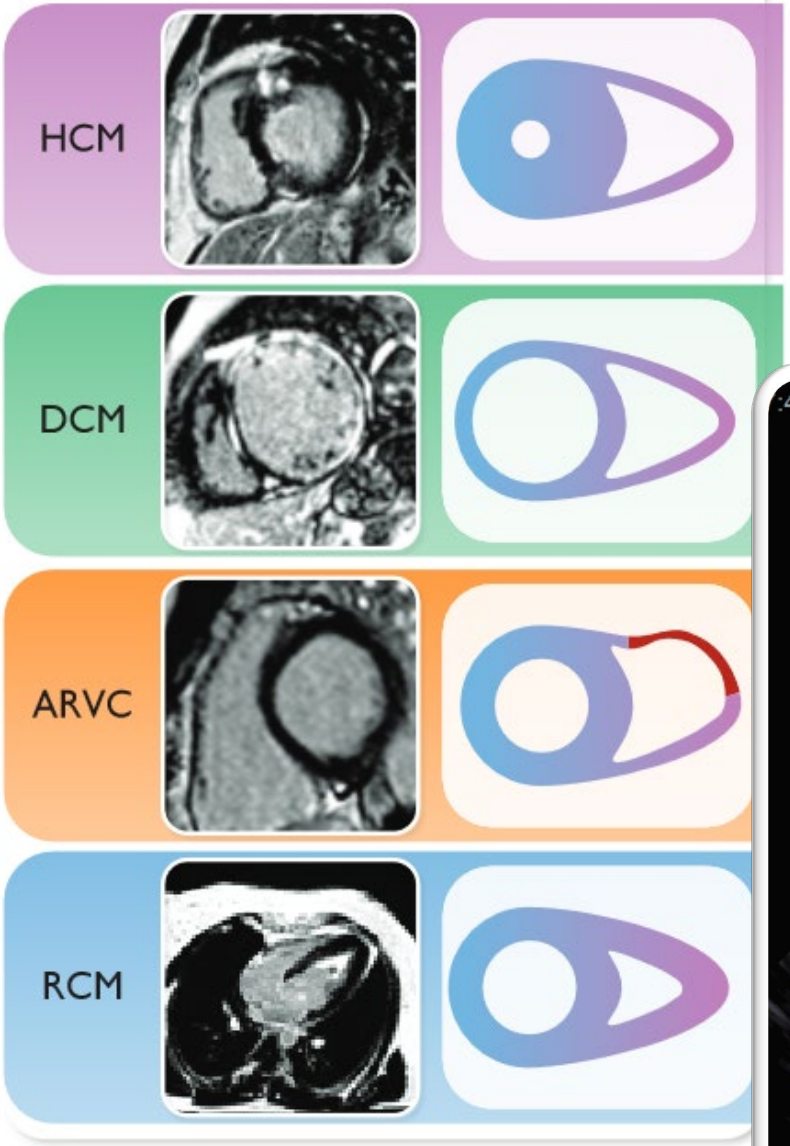


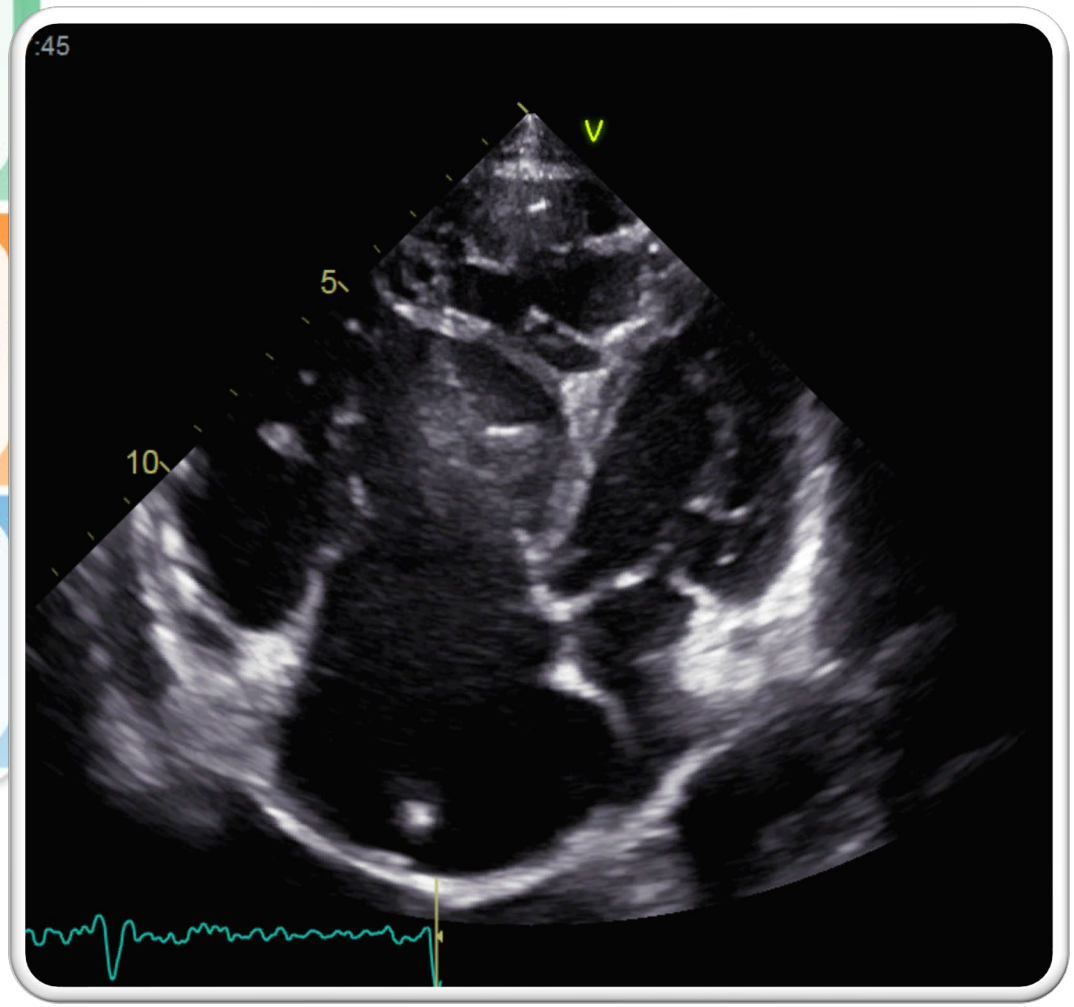
Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)



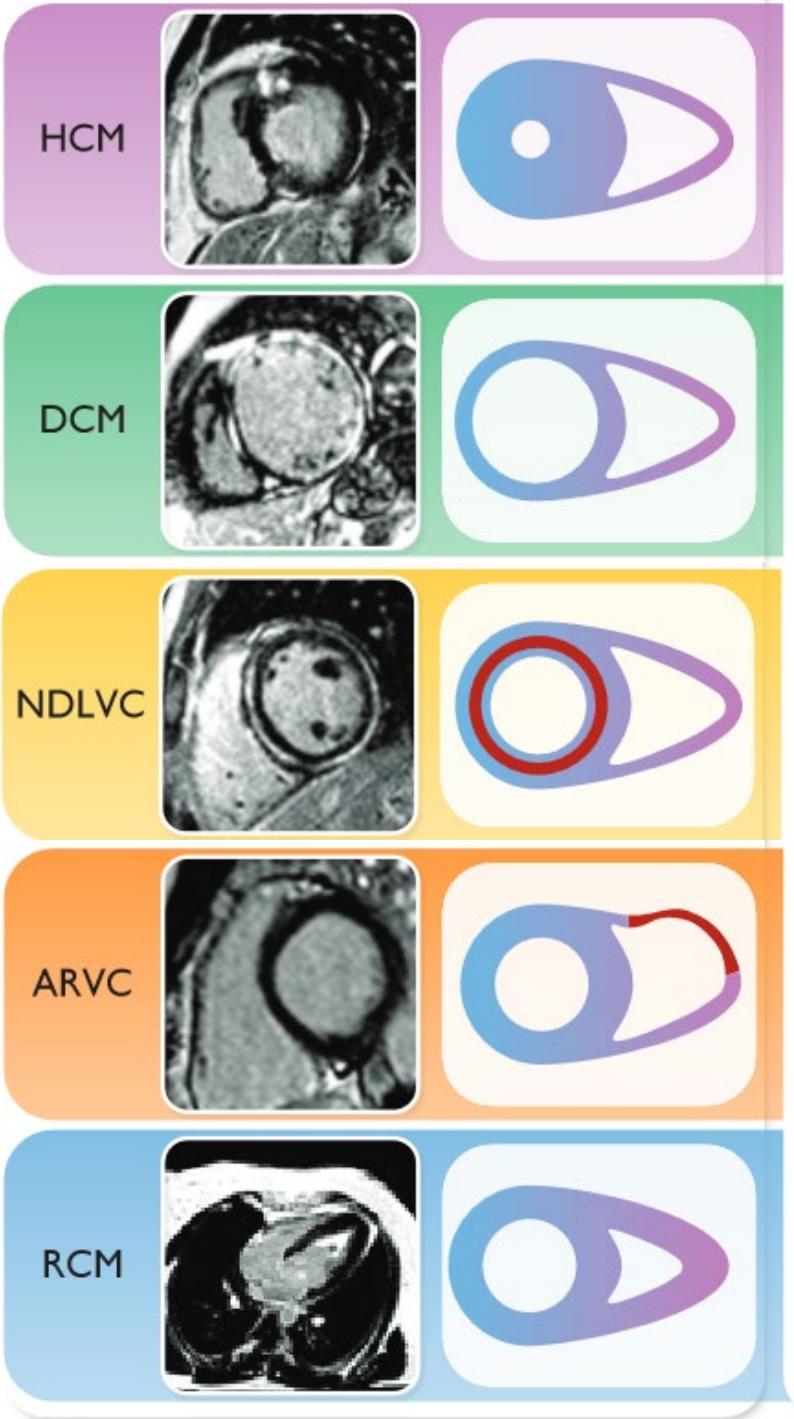


Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

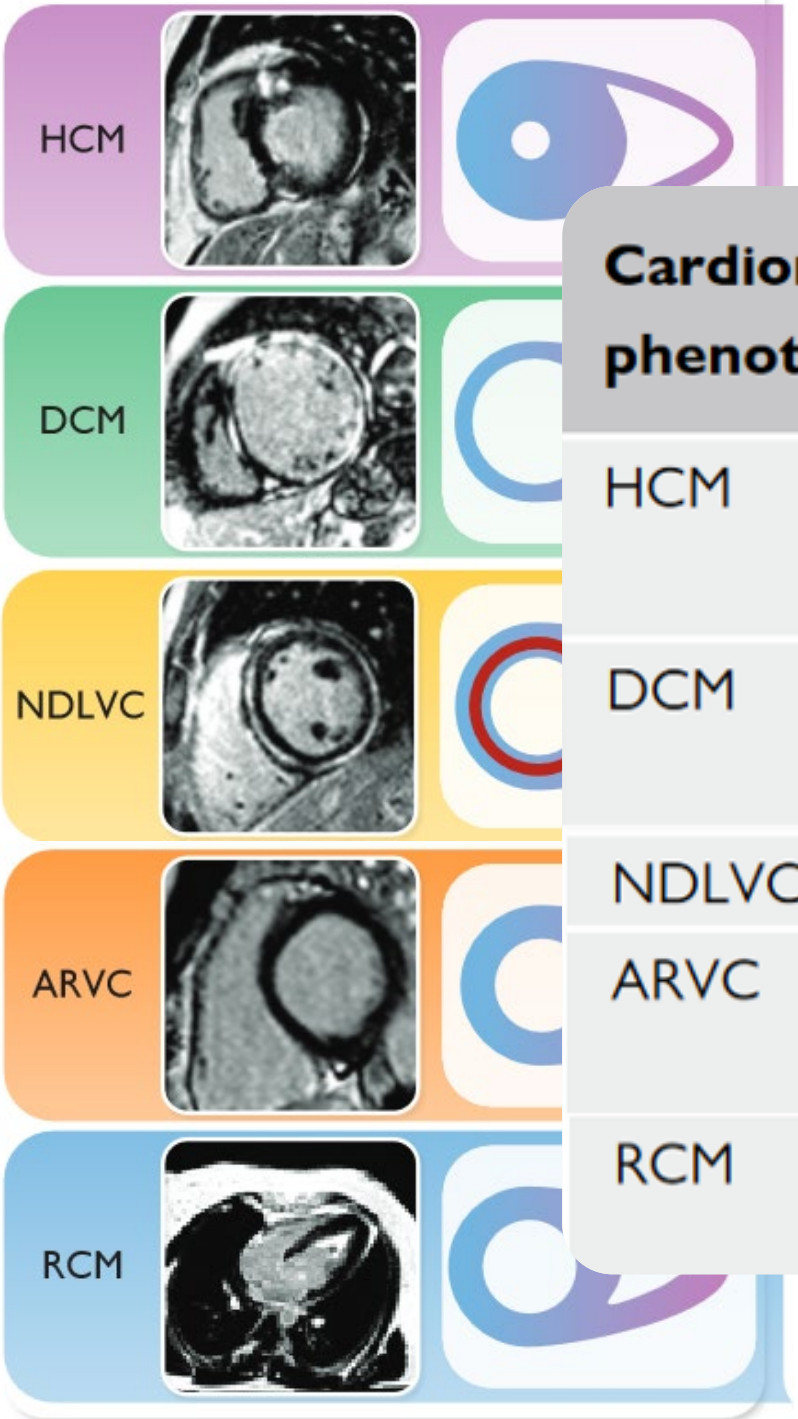




Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)



Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)



Cardiomyopathy phenotype	Adults
HCM	Prevalence: 0.2%[26–33]
DCM	Prevalence: 0.036–0.400%[25,37]
NDLVC	To be determined
ARVC	Prevalence: 0.078%[39–41]
RCM	Rare

Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

Diagnostik - allgemein

Diagnostik - allgemein

Recommendations	Class[a]	Level[b]
It is recommended that all patients with suspected or established cardiomyopathy undergo systematic evaluation using a multiparametric approach that includes clinical evaluation, pedigree analysis, ECG, Holter monitoring, laboratory tests, and multimodality imaging.[63]	I	C

Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

Diagnostik - allgemein

Recommendations	Class[a]	Level[b]
It is recommended that all patients with suspected or established cardiomyopathy undergo systematic evaluation using a multiparametric approach that includes clinical evaluation, pedigree analysis, ECG, Holter monitoring, laboratory tests, and multimodality imaging.[63]	I	C
It is recommended that all patients with suspected cardiomyopathy undergo evaluation of family history and that a three- to four-generation family tree is created to aid in diagnosis, provide clues to underlying aetiology, determine inheritance pattern, and identify at-risk relatives.[64–66]	I	C

Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

Diagnostik - allgemein

Recommendations	Class[a]	Level[b]
It is recommended that all patients with suspected or established cardiomyopathy undergo systematic evaluation using a multiparametric approach that includes clinical evaluation, pedigree analysis, ECG, Holter monitoring, laboratory tests, and multimodality imaging.[63]	I	C
It is recommended that all patients with suspected cardiomyopathy undergo evaluation of family history		
Genetic testing is recommended in patients fulfilling diagnostic criteria for cardiomyopathy in cases where it enables diagnosis, prognostication, therapeutic stratification, or reproductive management of the patient, or where it enables cascade genetic evaluation of their relatives who would otherwise be enrolled into long-term surveillance.[227–231,237,238]	I	B

Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

Diagnostik - Bildgebung

Diagnostik - Bildgebung

Recommendation Table 4 — Recommendation for echocardiographic evaluation in patients with cardiomyopathy

Recommendation	Class[a]	Level[b]
A comprehensive evaluation of cardiac dimensions and LV and RV systolic (global and regional) and LV diastolic function is recommended in all patients with cardiomyopathy at initial evaluation, and during follow-up, to monitor disease progression and aid risk stratification and management.[78,83–102]	I	B

Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

Diagnostik - Bildgebung

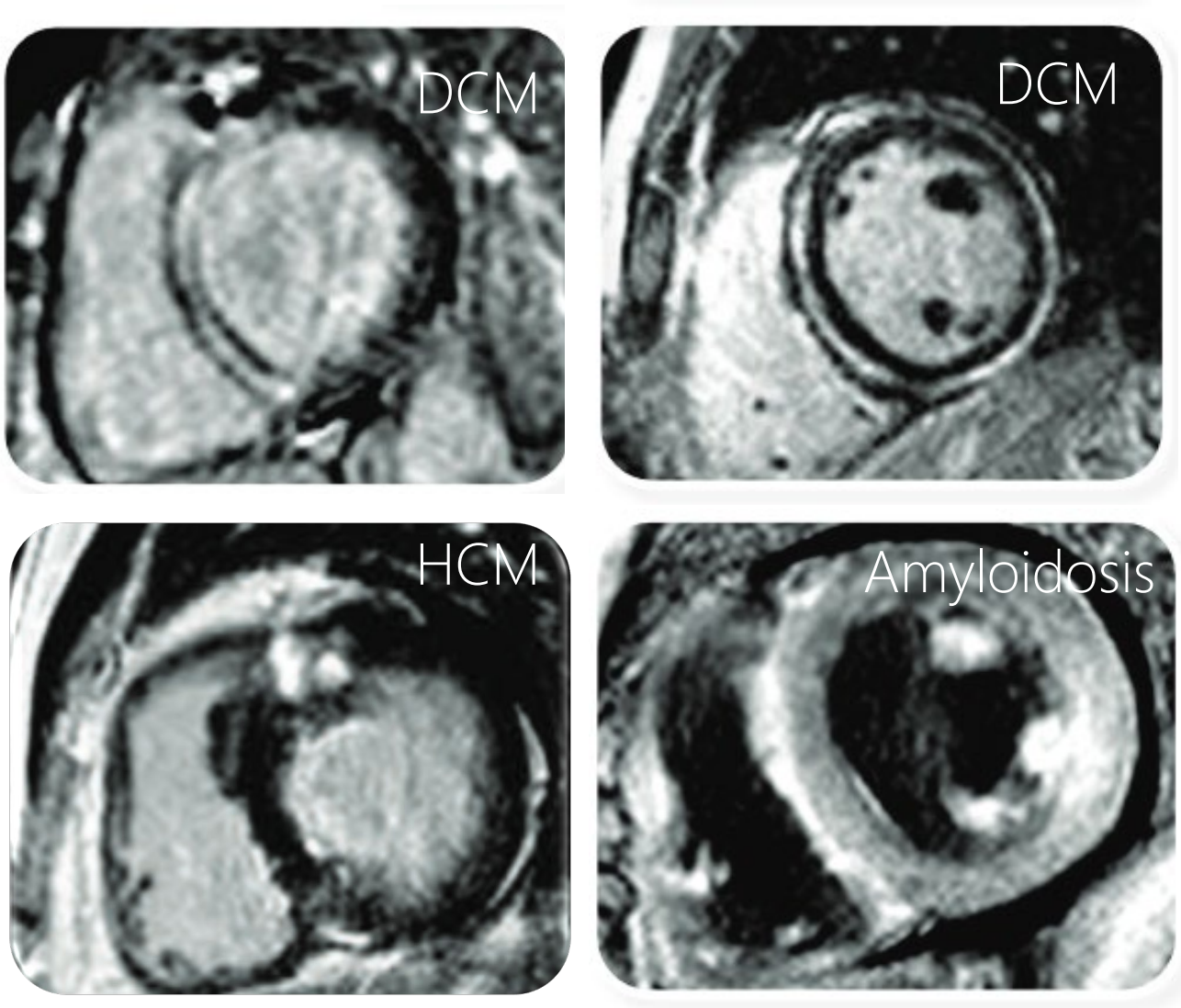
Recommendation Table 4 — Recommendation for echocardiographic evaluation in patients with cardiomyopathy

Recommendation	Class[a]	Level[b]
A comprehensive evaluation of cardiac dimensions and LV and RV systolic (global and regional) and LV diastolic function is recommended in all patients with cardiomyopathy at initial evaluation, and during follow-up, to monitor disease progression and aid risk stratification and management.[78,83–102]	I	B

Recommendations	Class[a]	Level[b]
Contrast-enhanced CMR is recommended in patients with cardiomyopathy at initial evaluation.[10,90,116,119–143]	I	B

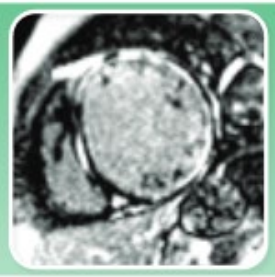
Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

Diagnostik - Bildgebung



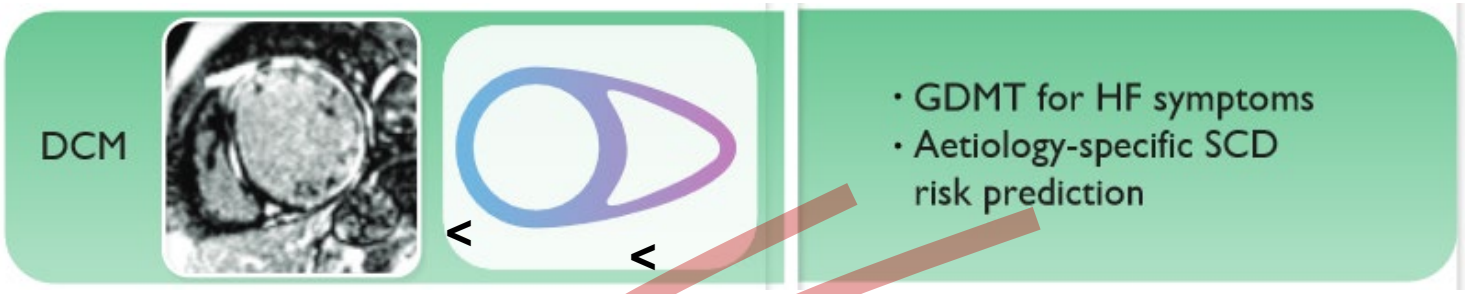
Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

DCM



- GDMT for HF symptoms
- Aetiology-specific SCD risk prediction

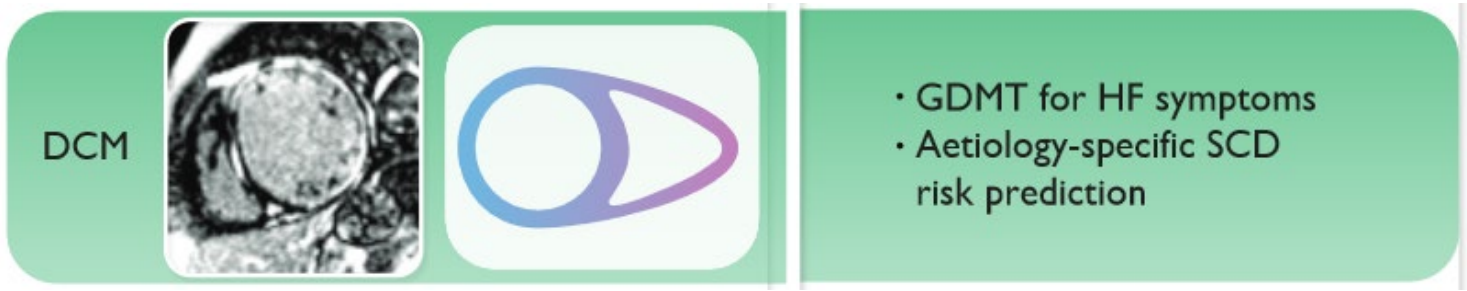
Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

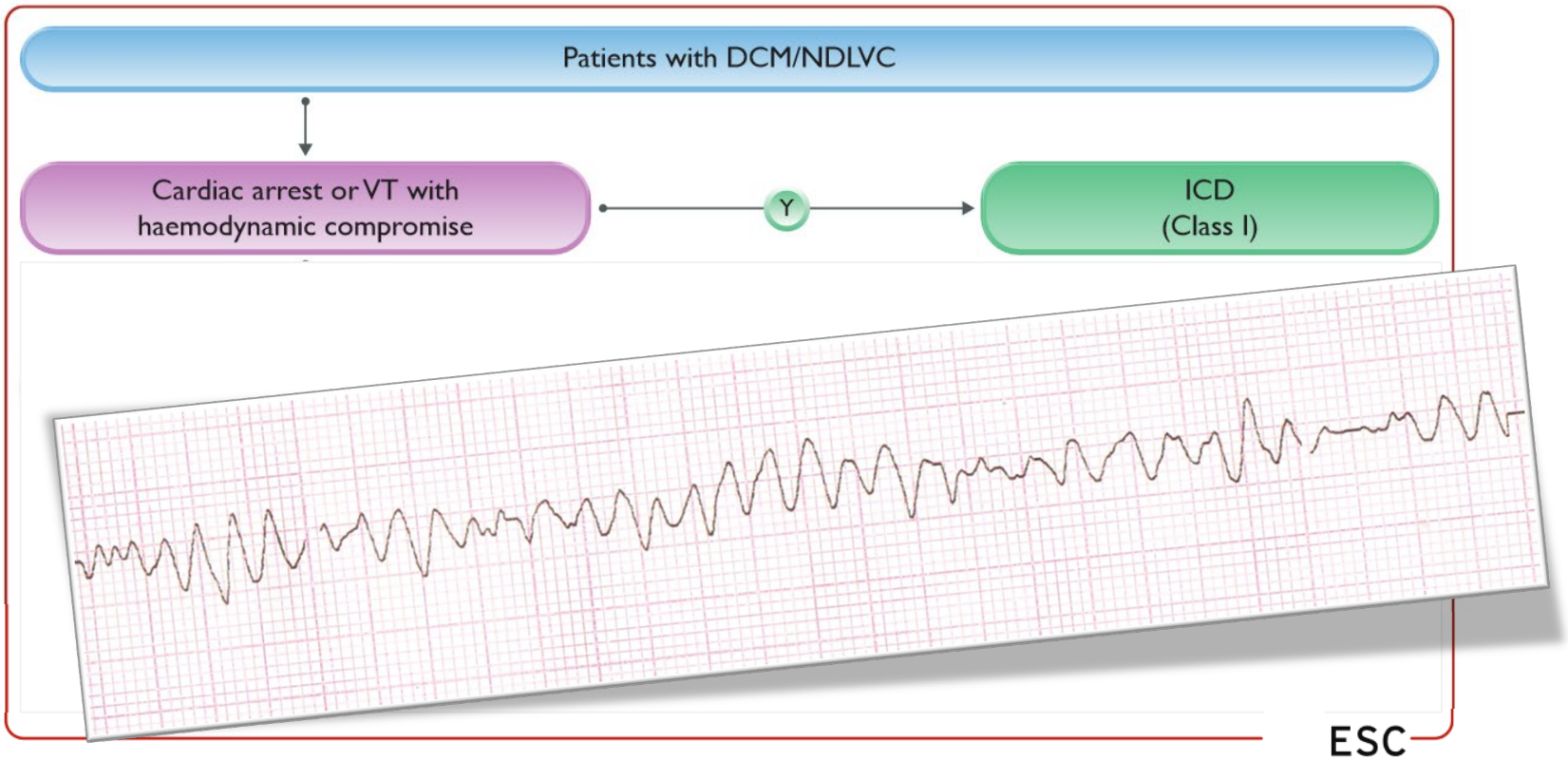


Symptome

Plötzlicher Herztod

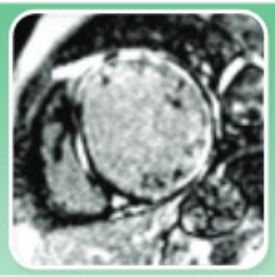
Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)





Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

DCM



- GDMT for HF symptoms
- Aetiology-specific SCD risk prediction

Patients with DCM/NDLVC

Cardiac arrest or VT with haemodynamic compromise

Y

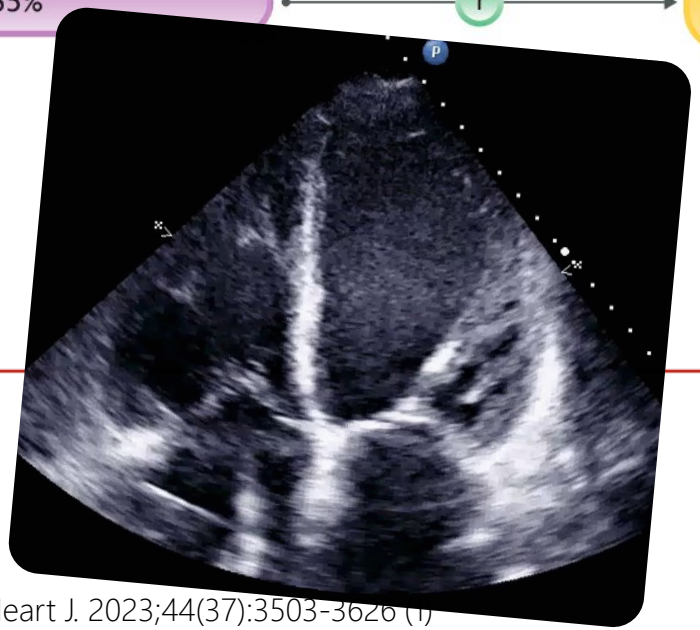
ICD (Class I)

N

LVEF <35%

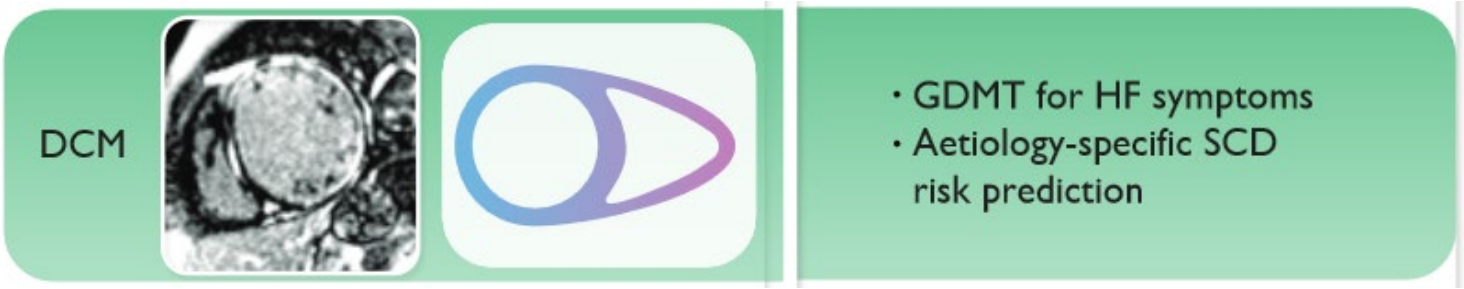
Y

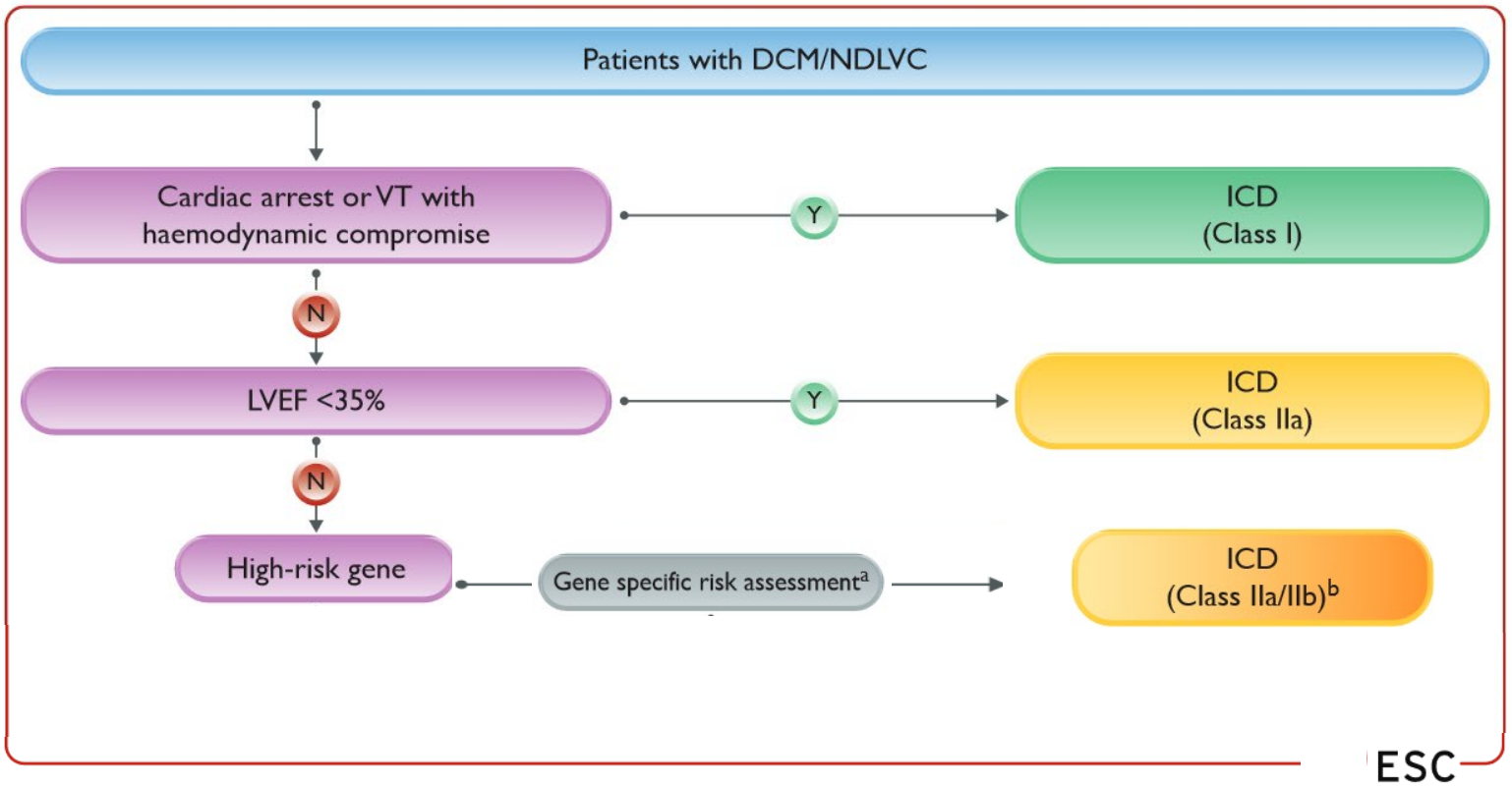
ICD (Class IIa)



ESC

Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)





Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

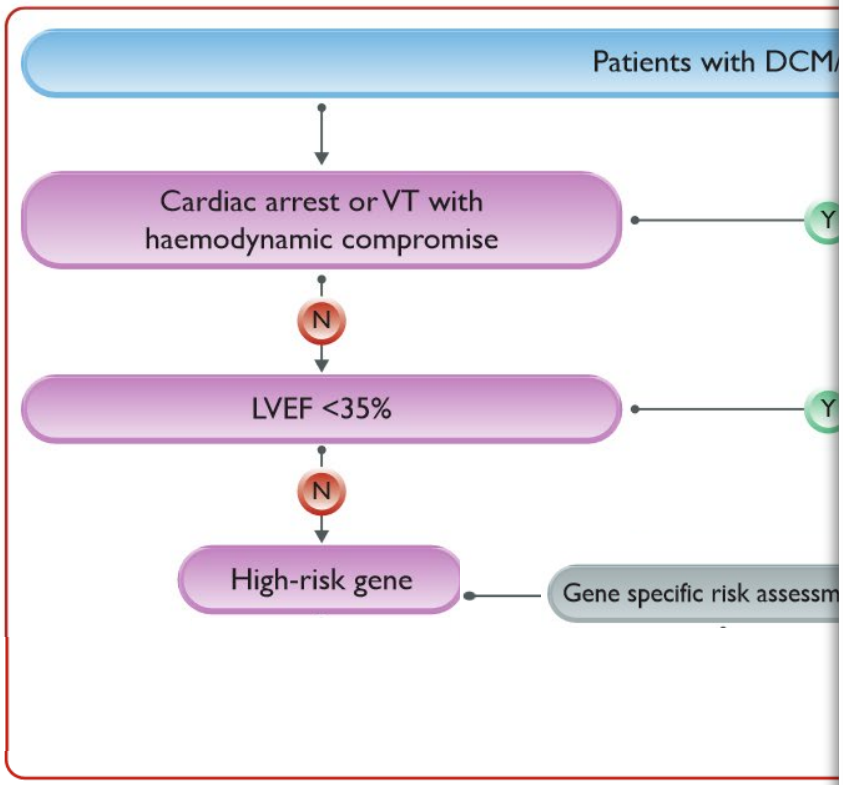


Table 21 High-risk genotypes and associated predictors of sudden cardiac death

Gene	Annual SCD rate	Predictors of SCD
LMNA[185,186,438,541,865,878,879]	5–10%	Estimated 5-year risk of life-threatening arrhythmia using *LMNA* risk score (https://lmna-risk-vta.fr)
FLNC-truncating variants[866,867,880]	5–10%	LGE on CMR LVEF < 45%
TMEM43[868,881]	5–10%	Male Female and any of the following: LVEF <45%, NSVT, LGE on CMR, >200 VE on 24h Holter ECG
PLN[542,882,883]	3–5%	Estimated 5-year risk of life-threatening arrhythmia using *PLN* risk score (https://plnriskcalculator.shinyapps.io/final_shiny) LVEF < 45% LGE on CMR NSVT
DSP[185,186]	3–5%	LGE on CMR LVEF < 45%
RBM20[869]	3–5%	LGE on CMR LVEF < 45%

Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

Table 21 High-risk genotypes and associated predictors of sudden cardiac death

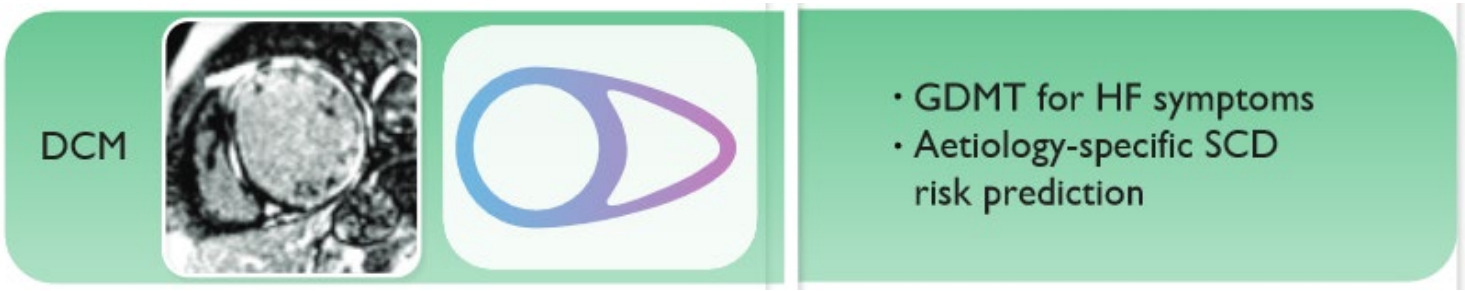
LMNA-risk VTA calculator

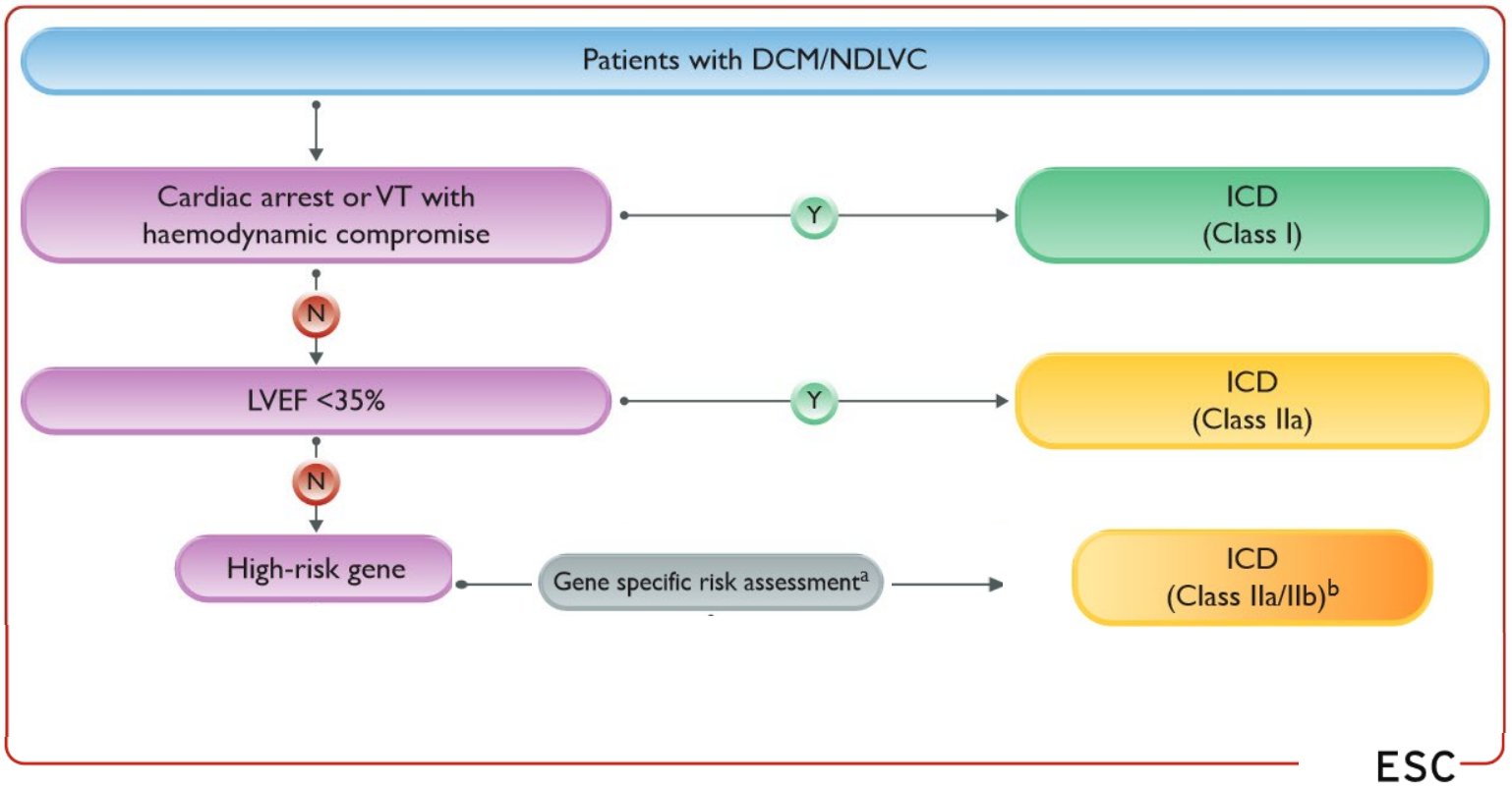
Risk Prediction Score for Life-Threatening Ventricular Tachyarrhythmias in Laminopathies

Sex	◉ Male ○ Female	
Non-missense LMNA mutation	○ Yes ◉ No	*Non-missense mutations include insertions, deletions, truncating mutations or mutations affecting splicing*
Atrio-ventricular block	○ Absent ◉ 1st degree ○ High degree	*Please select the highest degree. 1st degree AV block corresponds to ≥0.20 sec PR interval and high degree AV block to type II 2nd degree or 3rd degree (and not type I 2nd degree)*
Non-sustained ventricular tachycardia	◉ Yes ○ No	*NSVT corresponds to ≥3 consecutive ventricular complexes at a rate ≥120 bpm on 24-h ambulatory electrocardiographic monitoring*
Left ventricular ejection fraction	35 %	*Left ventricular ejection fraction measurement derived from echocardiogram*

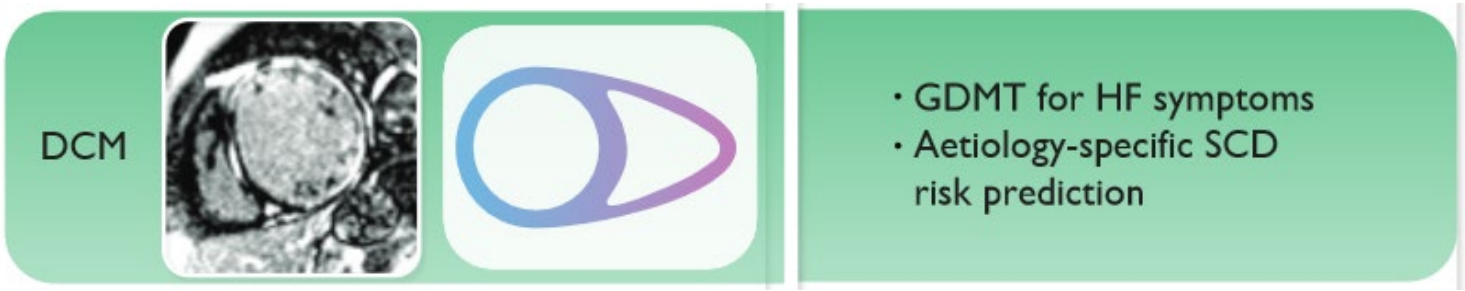
Risk of Life-Threatening Ventricular Tachyarrhythmias at 5 years
39.8 %

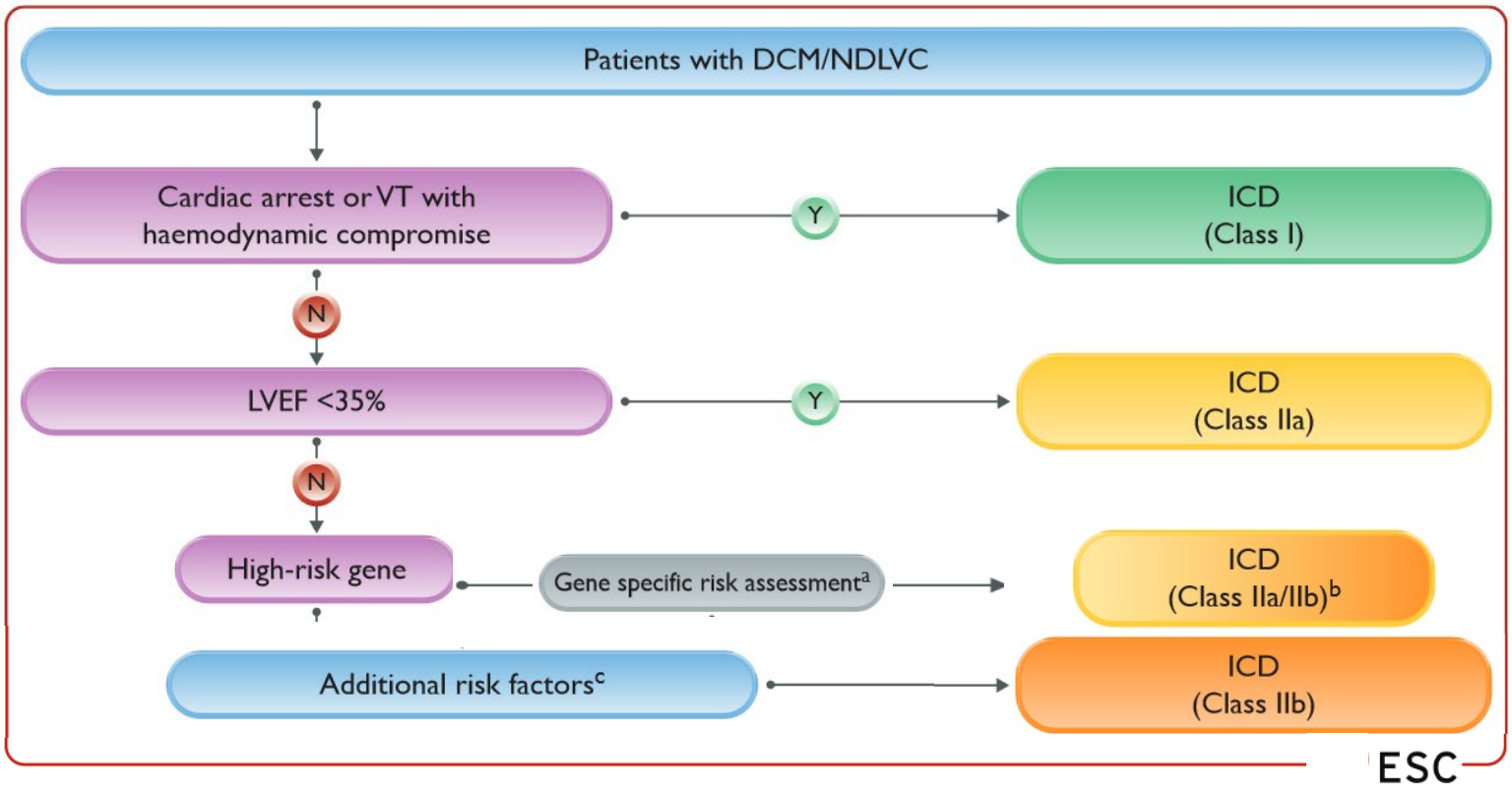
Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)





Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

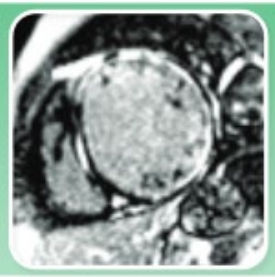


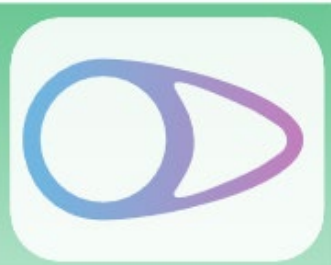


[c]Additional risk factors include syncope, LGE presence on CMR.

Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

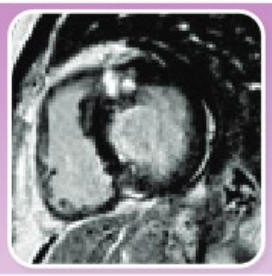
DCM





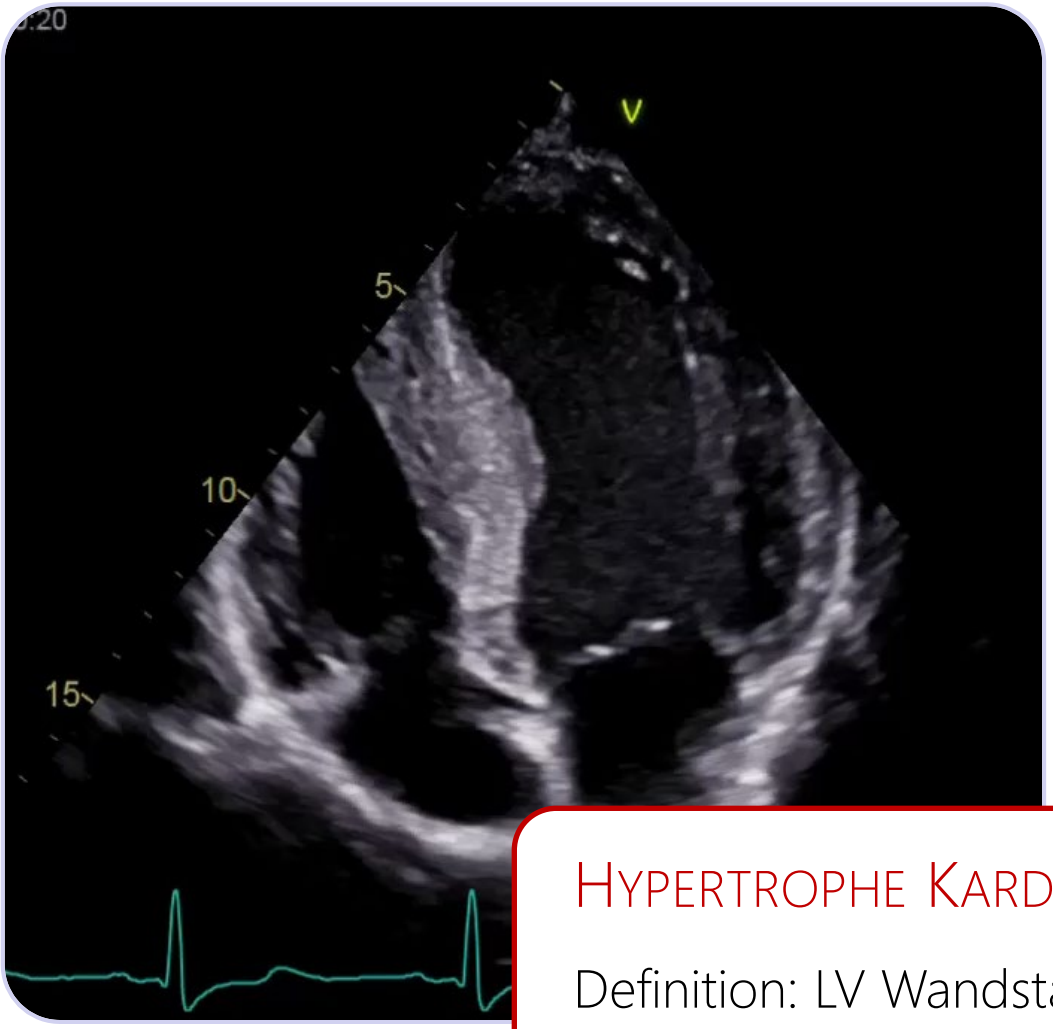
- GDMT for HF symptoms
- Aetiology-specific SCD risk prediction

HCM



- LVOTO management
- SCD risk prediction

Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)



Hypertrophe Kardiomyopathie

Definition: LV Wandstärke ≥ 15 mm in jedwedem Myokardsegment, nicht durch Nachlast erklärt.

Bei Verwandten 1° von HCM-Pat:: ≥ 13 mm

Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

“Sekundäre Ursache“ der HCM?

z. B.

Amyloidose

Anderson-Fabry´s Disease

Sarkoidose

Akute Myokarditis

Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

“Sekundäre Ursache“ der HCM?

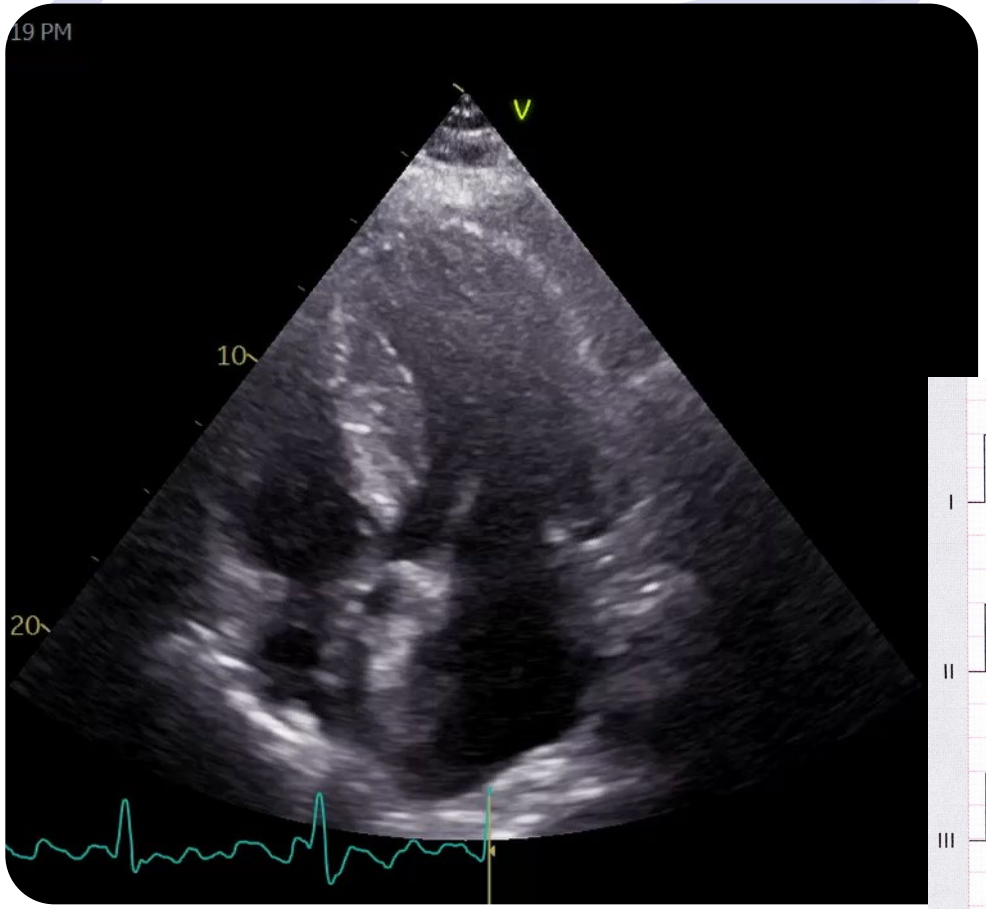
z. B.

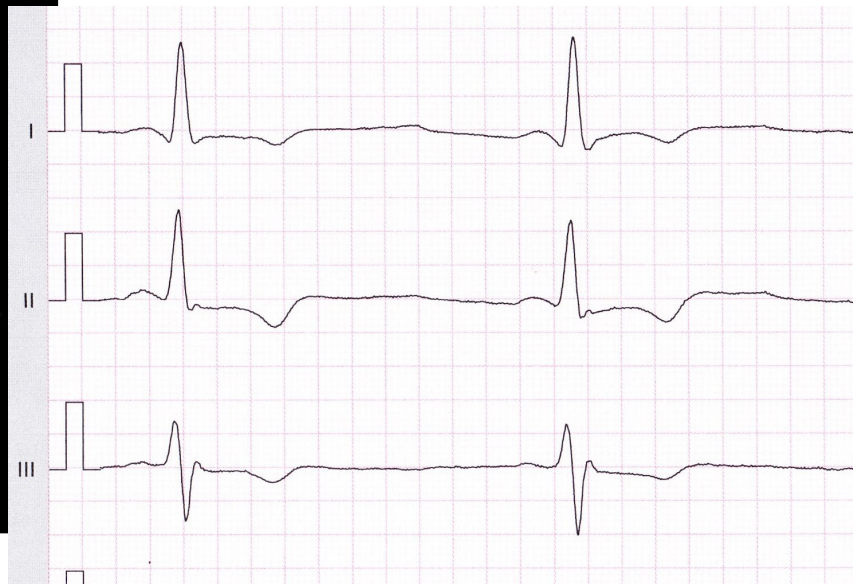
Amyloidose

Anderson-Fabry´s Disease

Sarkoidose

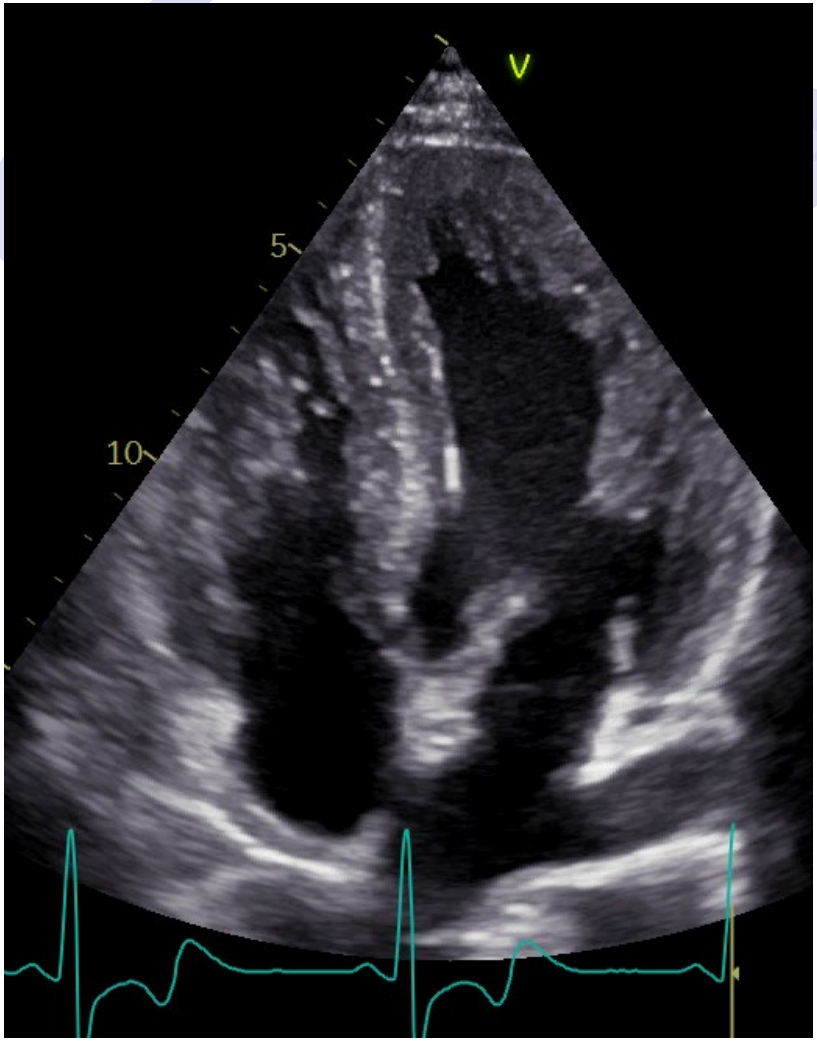
Akute Myokarditis





M Fabry

“Sekundäre Ursache“ der HCM?



M Fabry

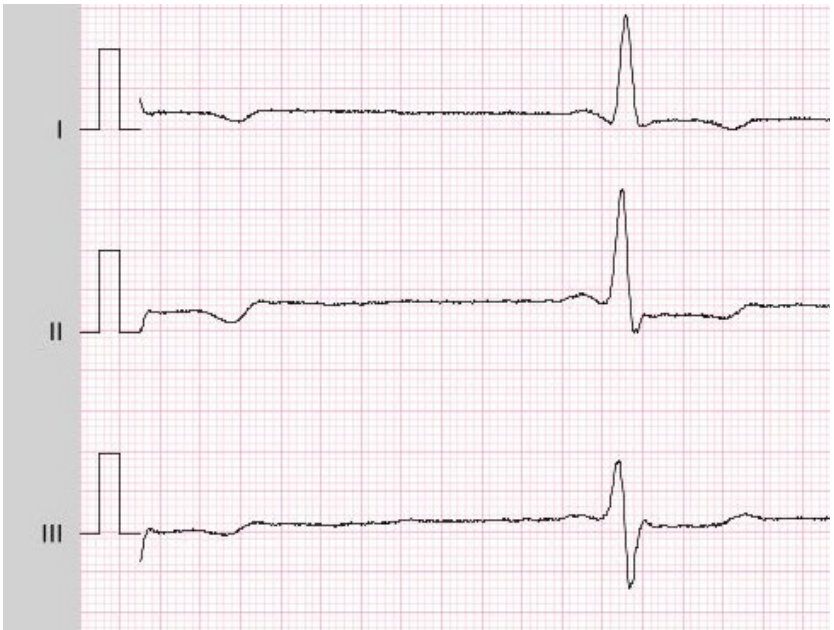
z. B.

Amyloidose

Anderson-Fabry´s Disease

Sarkoidose

Akute Myokarditis



“Sekundäre Ursache“ der HCM?

z. B.

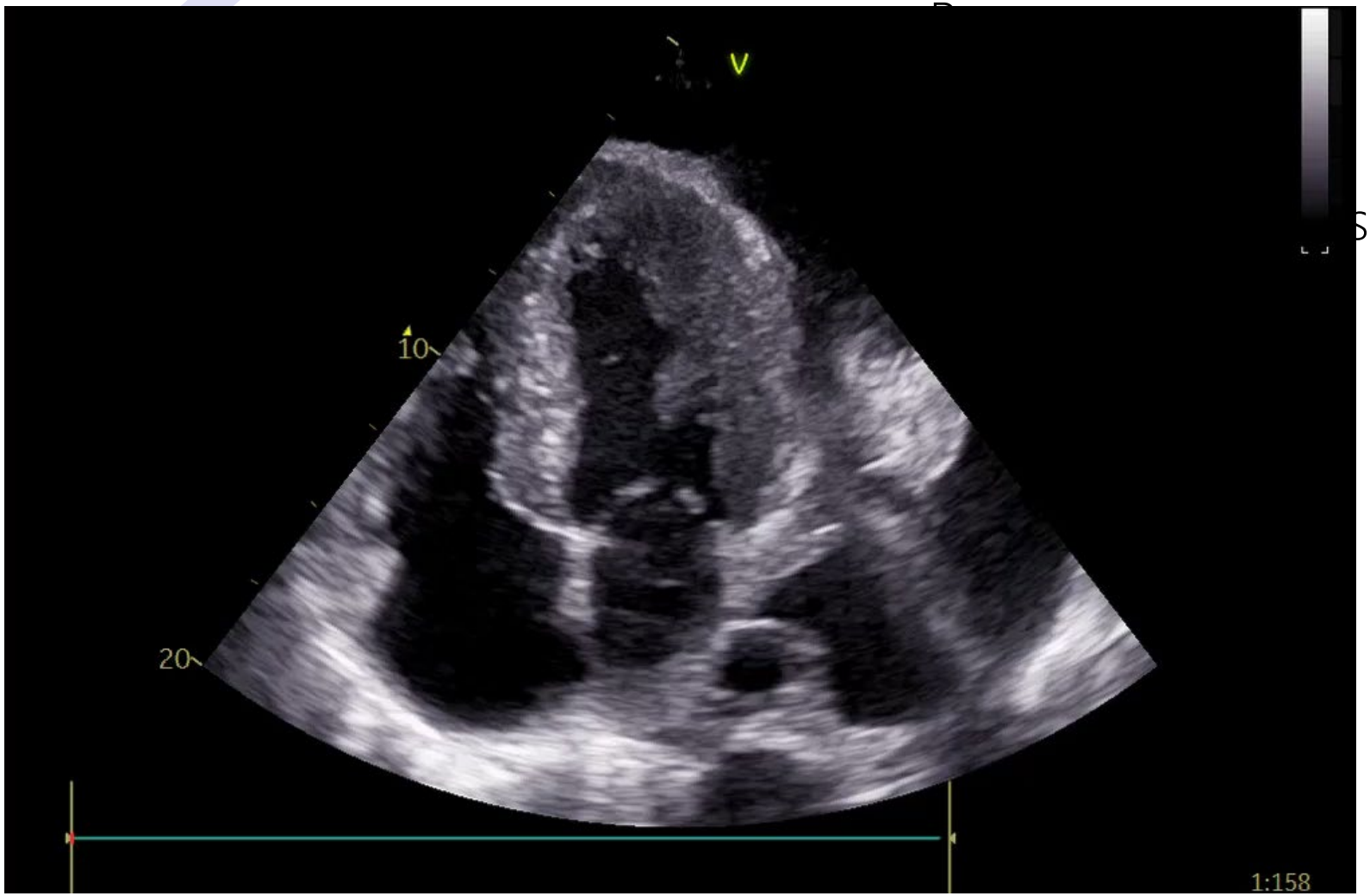
Amyloidose

Anderson-Fabry´s Disease

Sarkoidose

Akute Myokarditis

“Sekundäre Ursache“ der HCM?

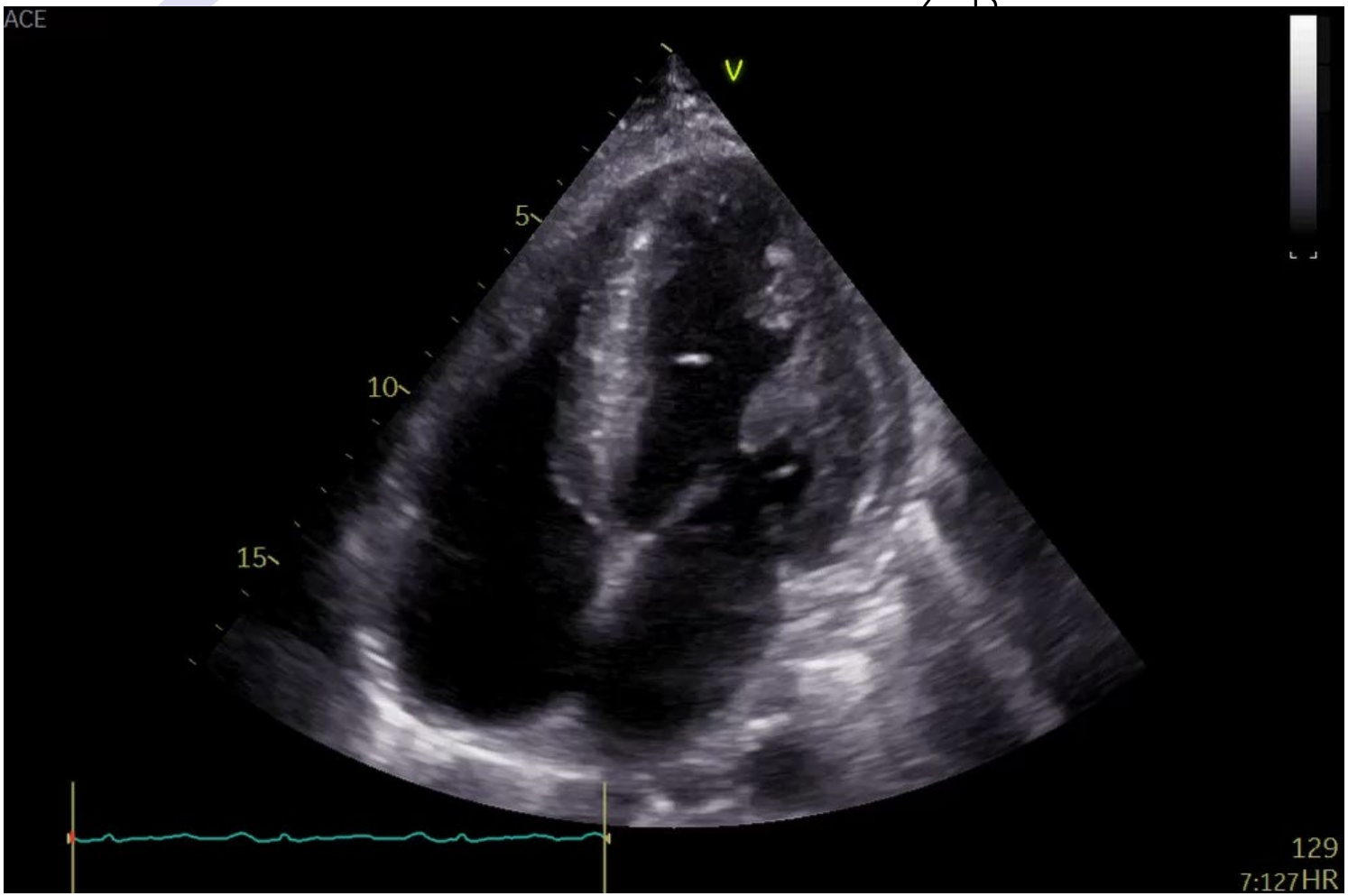


sease

Bakterielle Myokarditis

“Sekundäre Ursache“ der HCM?

z. B.



Disease

Bakterielle Myokarditis – 1 Woche zuvor

“Sekundäre Ursache“ der HCM?

z. B.

Amyloidose

Anderson-Fabry´s Disease

Akute Myokarditis

Sarkoidose

LVH ≥ 15 mm

LVH ≥ 13 mm (Verwandte 1°)

Hypertrophe Kardiomyopathie

Gradient ≥ 30 mmHg in Ruhe

Gradient ≥ 50 mmHg bei Belastung

Hypertroph-obstruktive Kardiomyopathie

Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

LVH ≥ 15 mm LVH ≥ 13 mm (Verwandte 1°)

Hypertrophe Kardiomyopathie

Gradient ≥ 30 mmHg in Ruhe
Gradient ≥ 50 mmHg bei Belastung

Hypertroph-obstruktive Kardiomyopathie

Berechnung des Risikos für plötzlichen Herztod: "HCM-Risk Calculator." ICD-Indikation entsprechend

Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

LVH ≥ 15 mm LVH ≥ 13 mm (Verwandte 1°)

Hypertrophe Kardiomyopathie

Gradient ≥ 30 mmHg in Ruhe
Gradient ≥ 50 mmHg bei Belastung

Hypertroph-obstruktive Kardiomyopathie

HCM Risk-SCD

Assess risk of sudden cardiac death and need for ICD in hypertrophic cardiomyopathy

Questions

1. **Age?**
2. Maximum LV Wall Thickness?
3. Left Atrial Size?
4. LVOT Gradient?
5. Family History of Sudden Cardiac Death?
6. Non-sustained VT?
7. Unexplained Syncope?

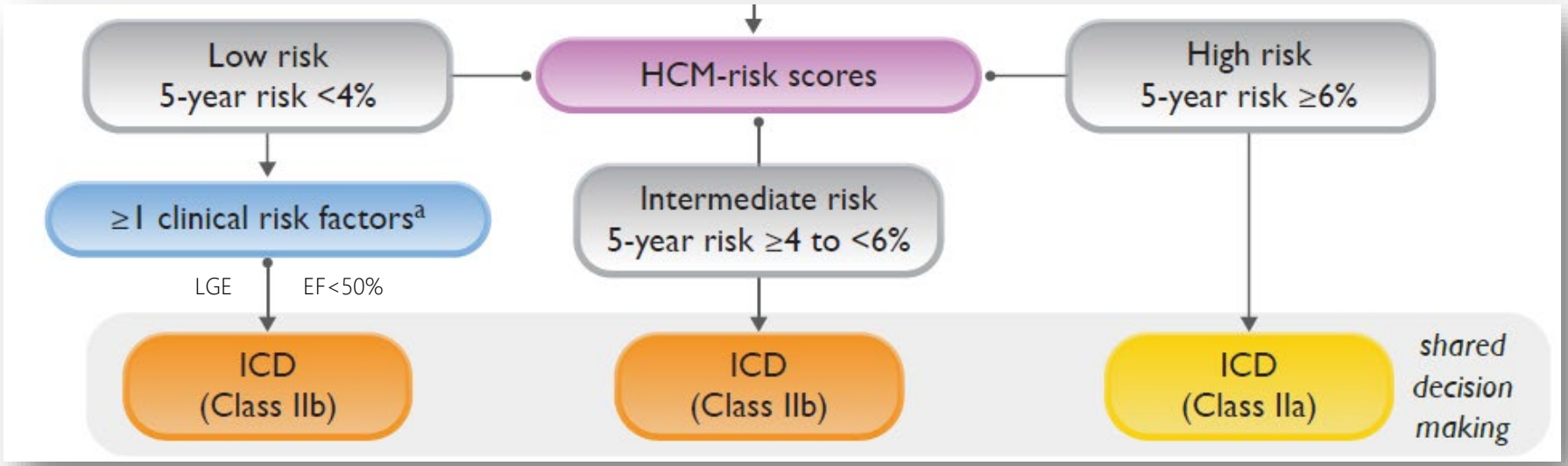
LVH ≥ 15 mm LVH ≥ 13 mm (Verwandte 1°)

Hypertrophe Kardiomyopathie

Gradient ≥ 30 mmHg in Ruhe
Gradient ≥ 50 mmHg bei Belastung

Hypertroph-obstruktive Kardiomyopathie

Berechnung des Risikos für plötzlichen Herztod: "HCM-Risk Calculator."



Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

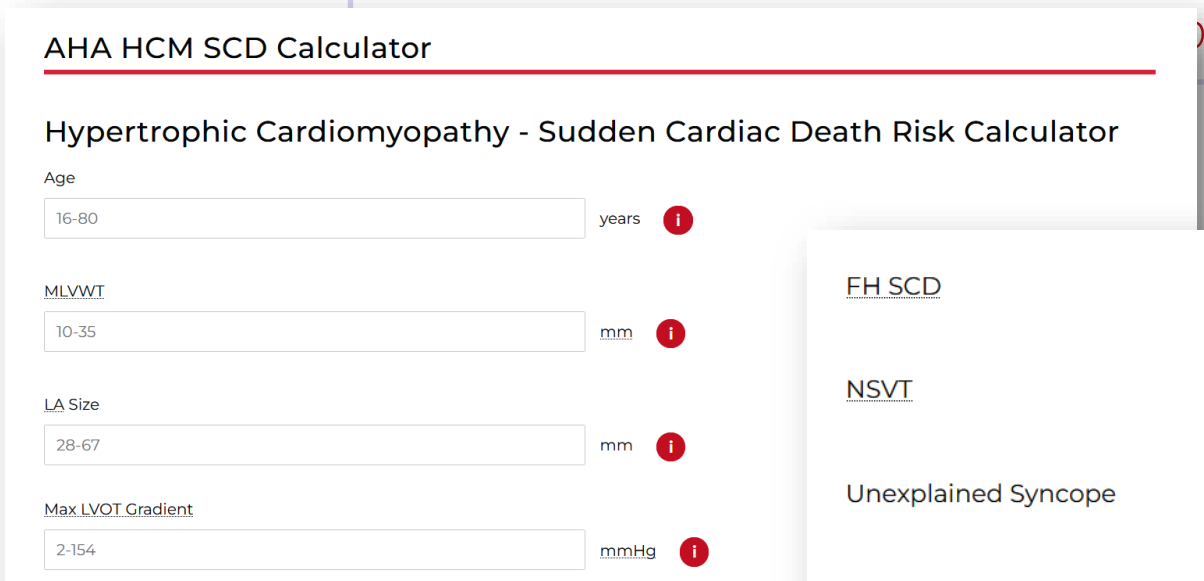
LVH ≥ 15 mm

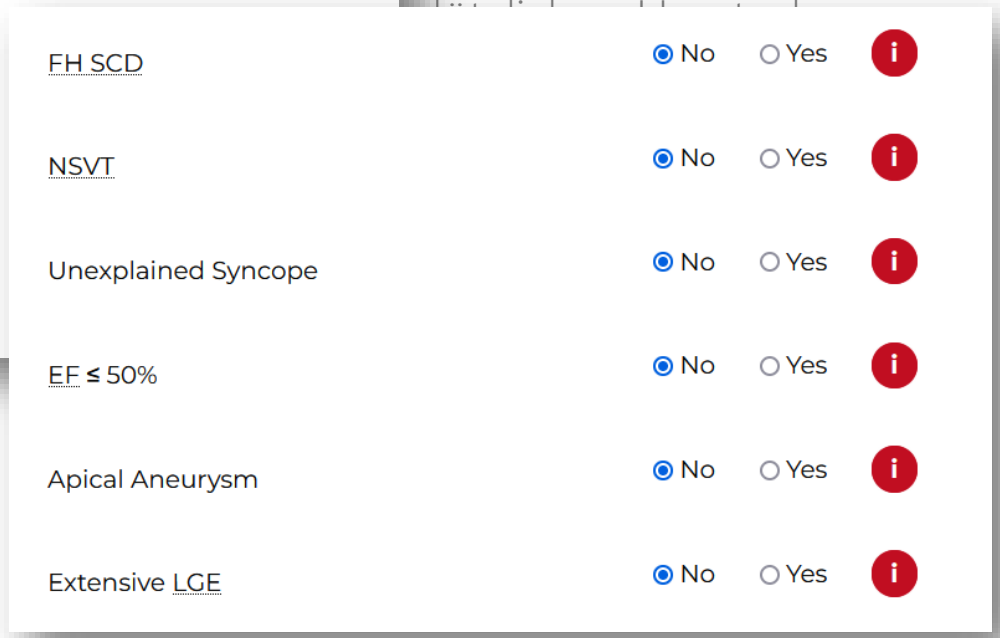
LVH ≥ 13 mm (Verwandte 1°)

Hypertrophe Kardiomyopathie

Gradient ≥ 30 mmHg in Ruhe

Gradient ≥ 50 mmHg bei Belastung



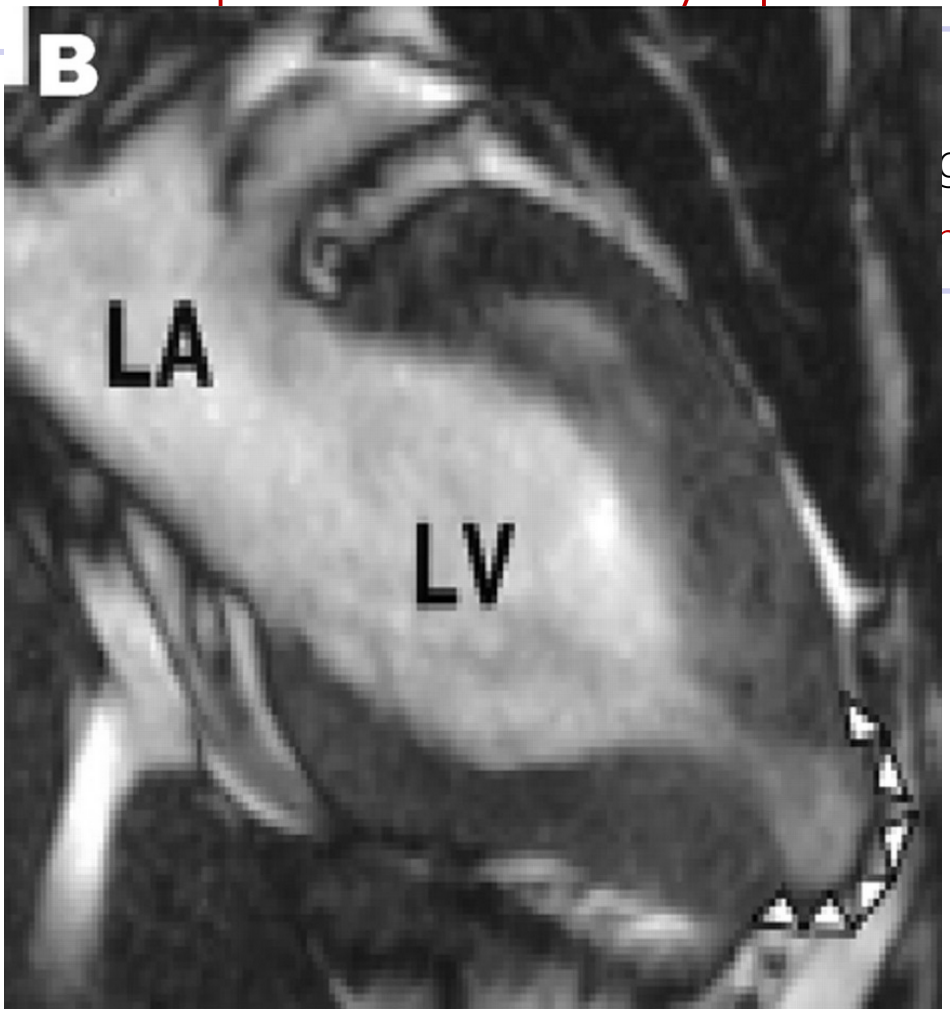


Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

LVH ≥ 15 mm

LVH ≥ 13 mm (Verwandte 1°)

Hypertrophe Kardiomyopathie



g

myopathie

Berechnung des Risikos für plötzlichen Herztod: "HCM-Risk Calculator." ICD-Indikation entsprechend

Maron MS et al, Circulation. 200;118(15):1541-9 (2)

LVH ≥ 15 mm LVH ≥ 13 mm (Verwandte 1°)

Hypertrophe Kardiomyopathie

Gradient ≥ 30 mmHg in Ruhe
Gradient ≥ 50 mmHg bei Belastung

Hypertroph-obstruktive Kardiomyopathie

Berechnung des Risikos für plötzlichen Herztod: "HCM-Risk Calculator." ICD-Indikation entsprechend

Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

LVH ≥ 15 mm LVH ≥ 13 mm (Verwandte 1°)

Hypertrophe Kardiomyopathie

Gradient ≥ 30 mmHg in Ruhe
Gradient ≥ 50 mmHg bei Belastung

Hypertroph-obstruktive Kardiomyopathie

"Outflow tract Management" (≥ 50 mmHg)

Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

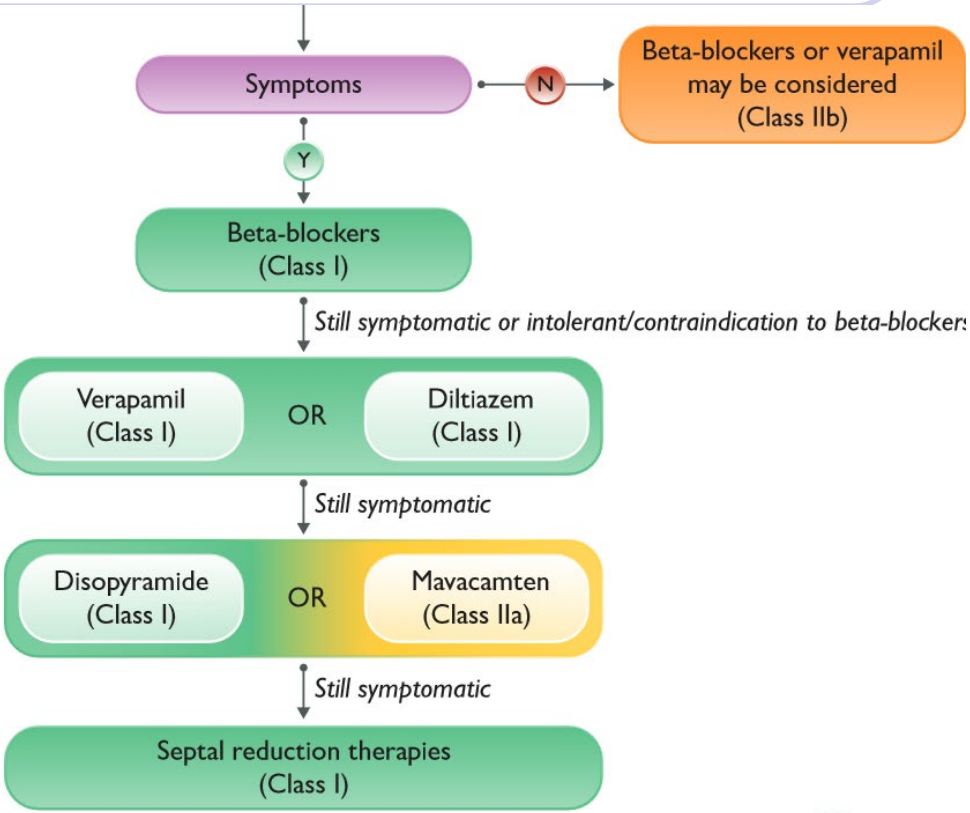
LVH ≥ 15 mm LVH ≥ 13 mm (Verwandte 1°)

Hypertrophe Kardiomyopathie

Gradient ≥ 30 mmHg in Ruhe

Gradient ≥ 50 mmHg bei Belastung

Hypertroph-obstruktive Kardiomyopathie



Arbelo E et al, Eur Heart J. 2023; 44(37):3503-3626 (1)

LVH ≥ 15 mm LVH ≥ 13 mm (Verwandte 1°)

Hypertrophe Kardiomyopathie

Gradient ≥ 30 mmHg in Ruhe
Gradient ≥ 50 mmHg bei Belastung

Hypertroph-obstruktive Kardiomyopathie

Kein Digitalis, keine Nitrate, PDA Inhibitoren

↓ Symptome, Gradient ≥ 50 mmHg

Beta Blocker (I)

↓ Intolerant/ Noch Symptome

Verapamil (I) oder Diltiazem (I)

↓ Noch Symptome

Disopyramid (I) oder Mavacamten (IIa)

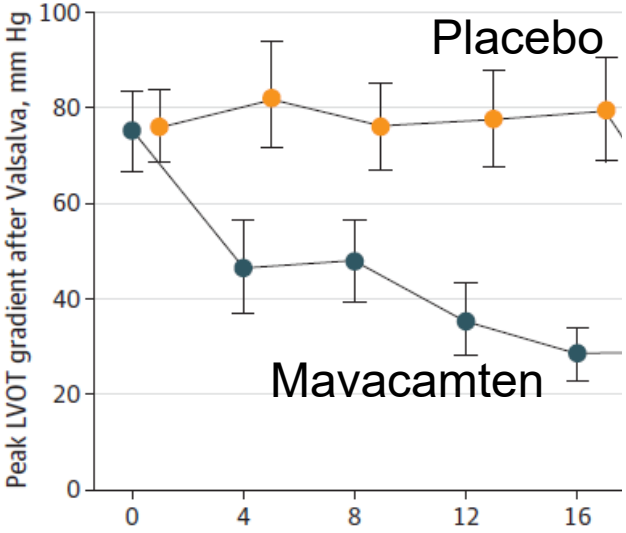
↓ Noch Symptome

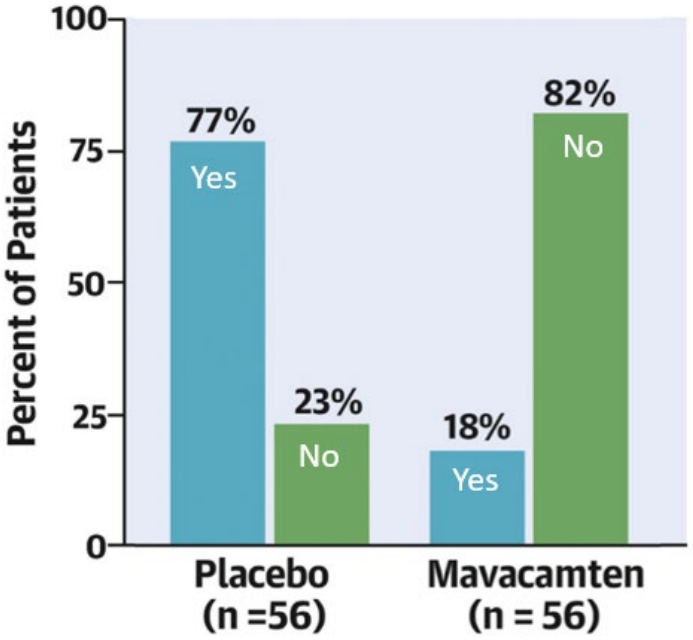
Septale Reduktionstherapie (Myektomie oder TASH) (I)

Arbelo E et al, Eur Heart J. 2023;44(37):3503-3626 (1)

Mevacamten, VALOR-HCM Studie

Peak LVOT Gradient, Valsalva

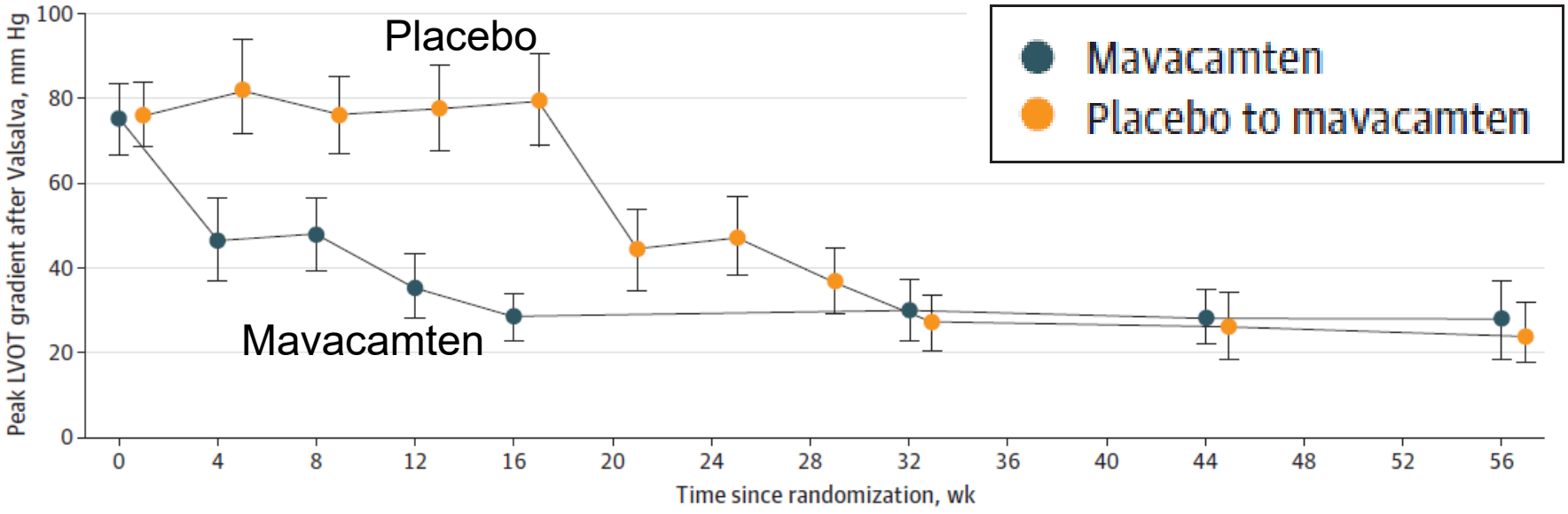




Desai M, et al. J Am Coll Cardiol. 2022;80:95-108 (3)

Mevacamten, VALOR-HCM Studie

Peak LVOT Gradient, Valsalva

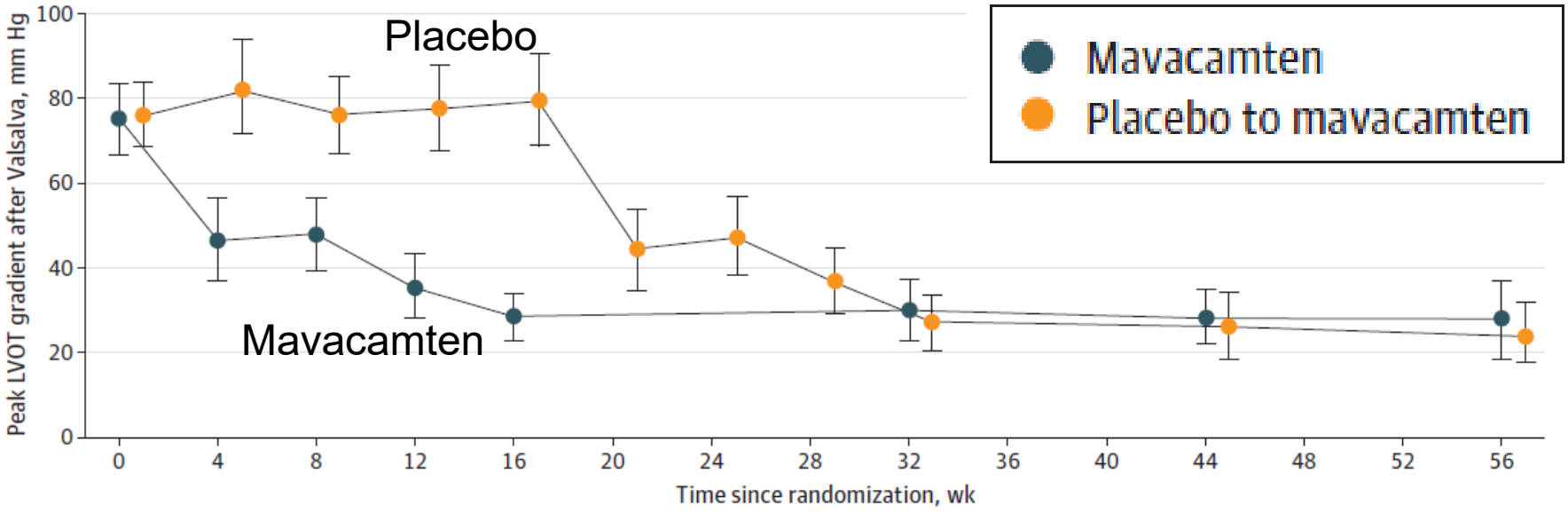


Desai M, et al. J Am Coll Cardiol. 2022;80:95-108 (3)

Desai M, et al. JAMA Cardiol. 202;8(10):968-977 (4)

Mevacamten, VALOR-HCM Studie

Peak LVOT Gradient, Valsalva



12/108: Dosisanpassung/Unterbrechung

Desai M, et al. J Am Coll Cardiol. 2022;80:95-108 (3)

Desai M, et al. JAMA Cardiol. 202;8(10):968-977 (4)

Therapie mit Mevacamten

LVOT Gradient ≥ 50 mmHg

NHYA II-III trotz med. Therapie

LVEF ≥ 55%

Cave: embryofetale Toxizität, vor Beginn neg. Schwangerschaftstest, sichere Verhütung bis 6 Monate nach letzter Einnahme

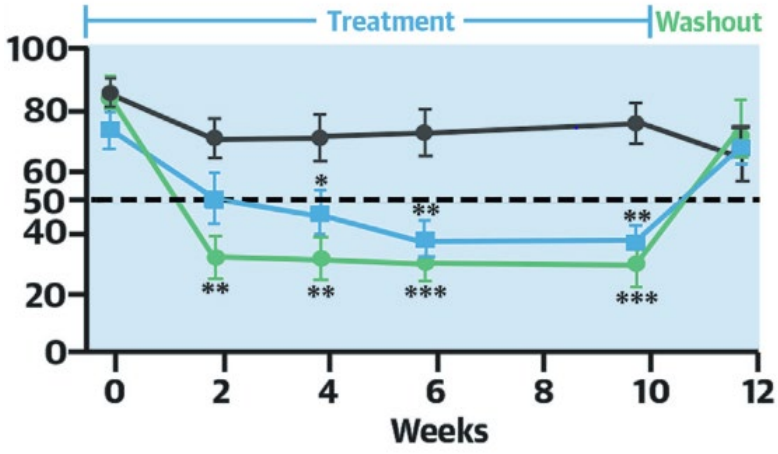
Bestimmung des CYP2C19 Metabolismus

Startdosis 2.5 – 5 mg/d je nach Metabolismus

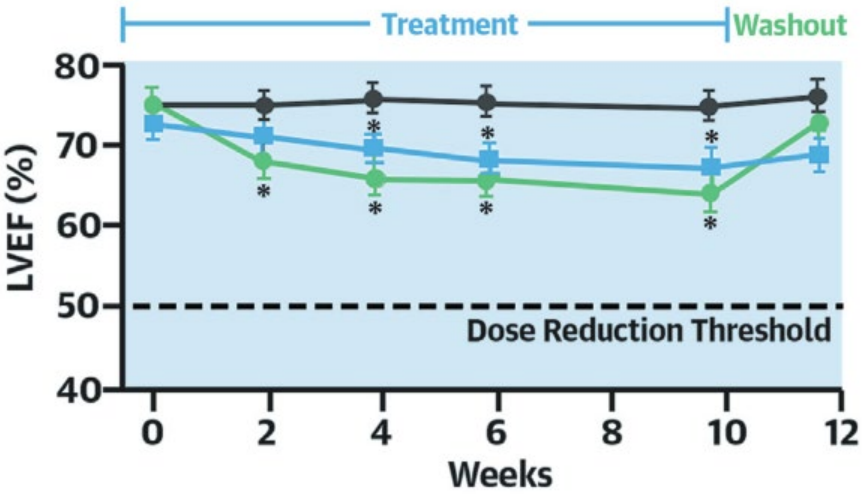
Steigerung oder Reduktion je nach LVOT Gradient und EF (< 50% => Stop)

Aficamten, “Redwood HCM“

Peak LVOT Gradient, Valsalva (mmHg)



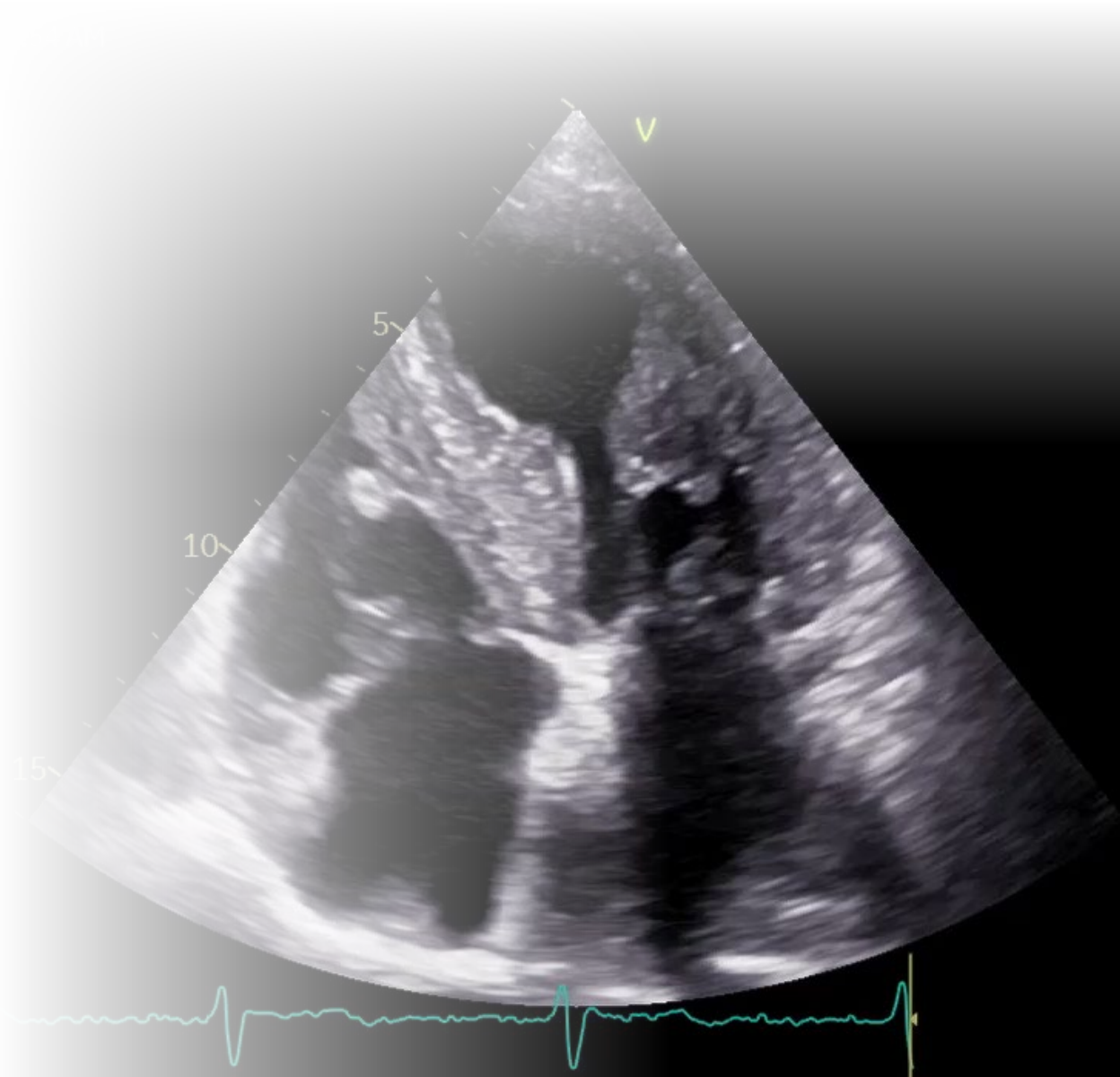
Ejection Fraction (%)



Pooled placebo group (n = 13)
Aficamten cohort 1 (n = 14)
Aficamten cohort 2 (n = 14)

Maron MS, et al. J Am Coll Cardiol 2023;81:34–45 (5)

Kardiale Amyloidose

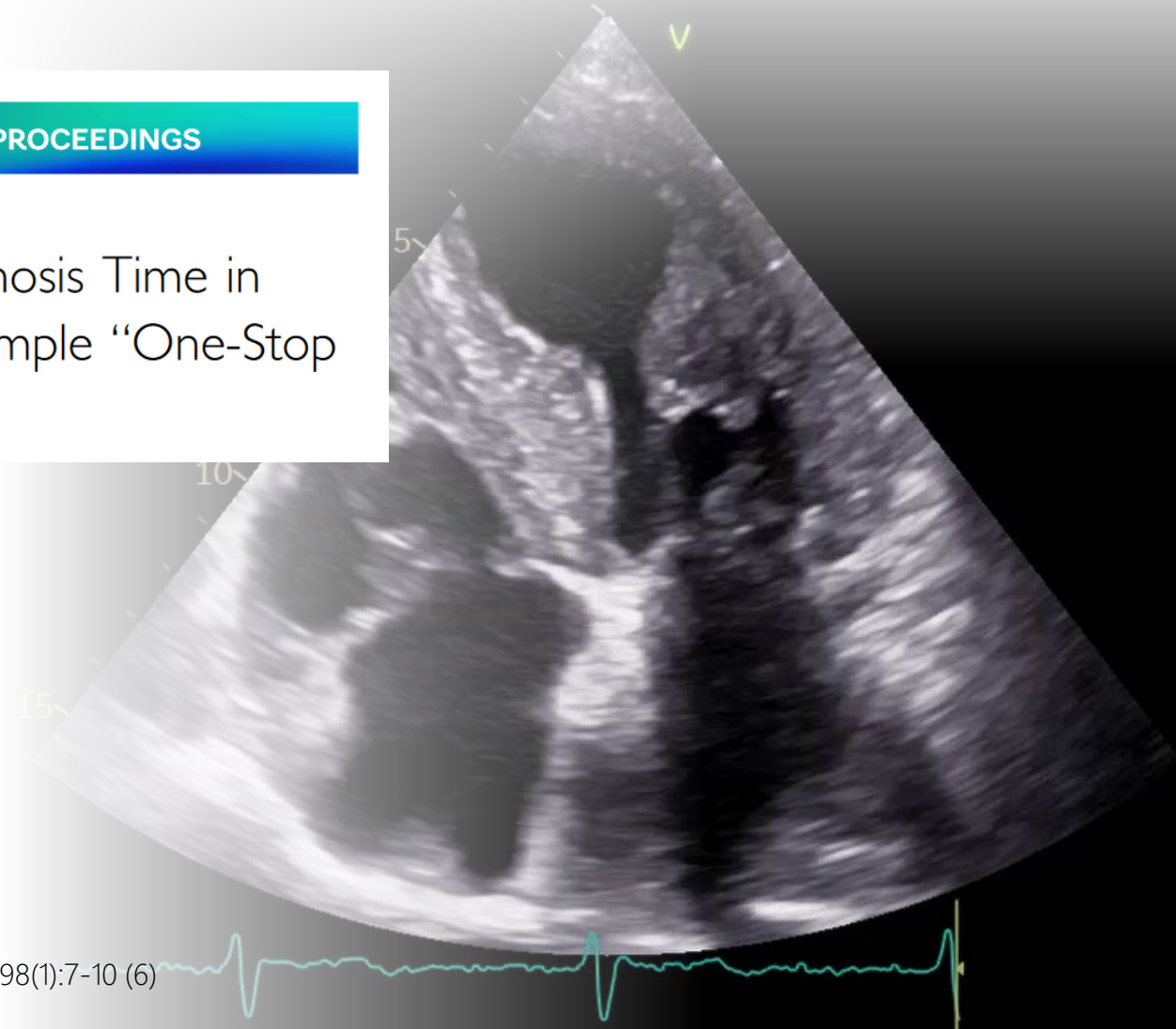


Kardiale Amyloidose

MAYO CLINIC

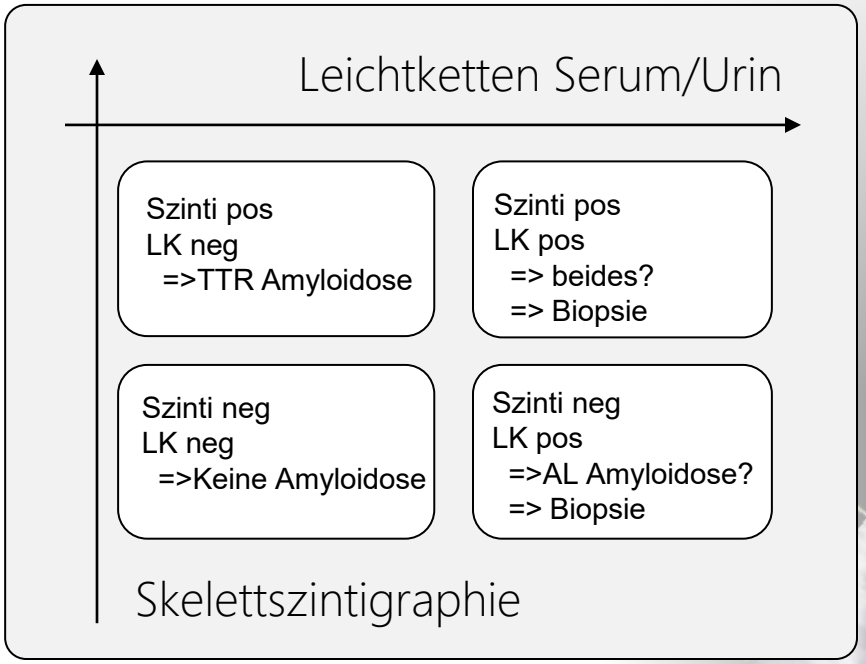
MAYO CLINIC PROCEEDINGS

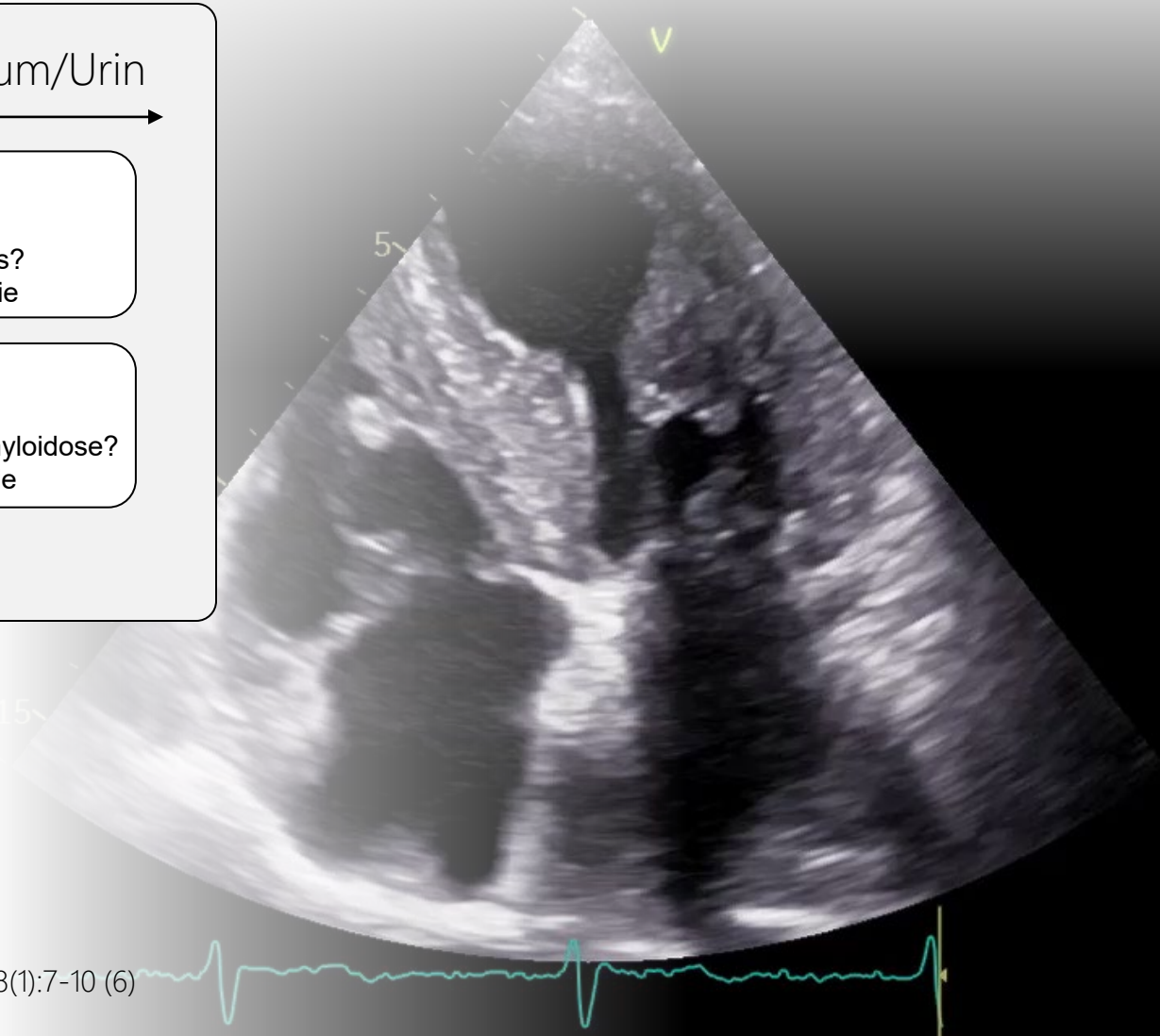
Decreasing Door-to-Diagnosis Time in Cardiac Amyloidosis: A Simple "One-Stop Shop" Approach



Grogan M, et al. Mayo Clin Proc. 2023 Jan;98(1):7-10 (6)

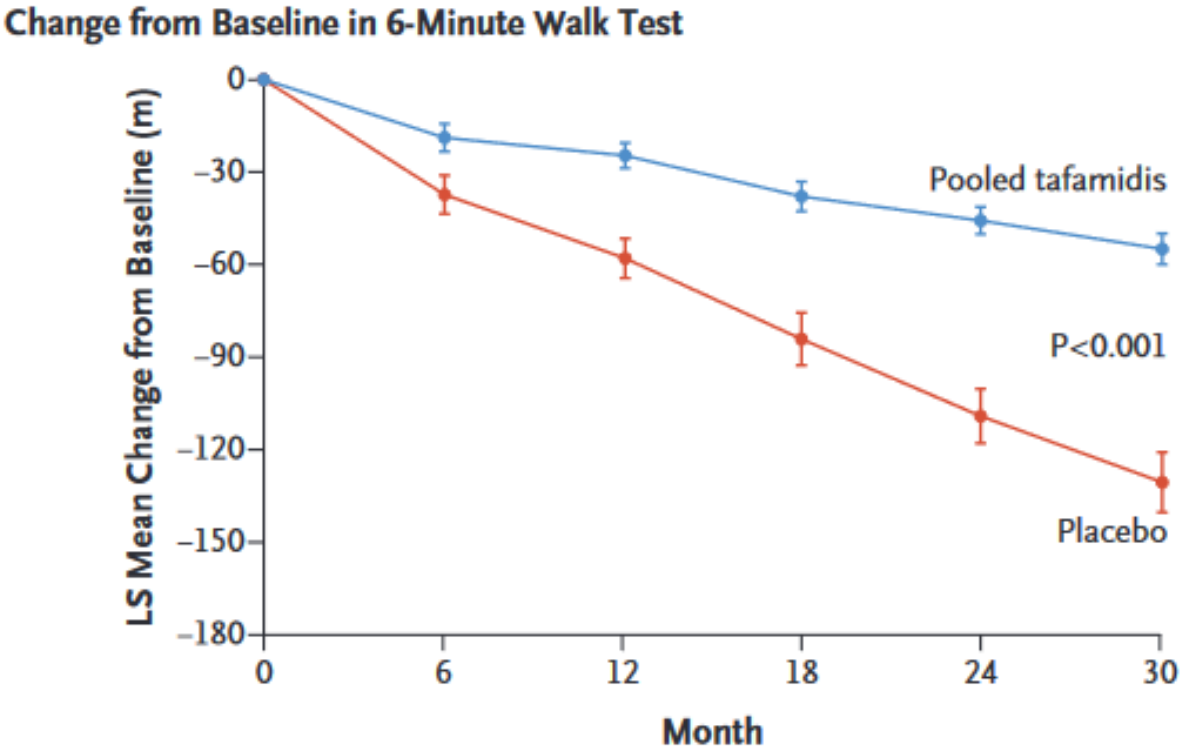
Kardiale Amyloidose





Grogan M, et al. Mayo Clin Proc. 2023 Jan;98(1):7-10 (6)

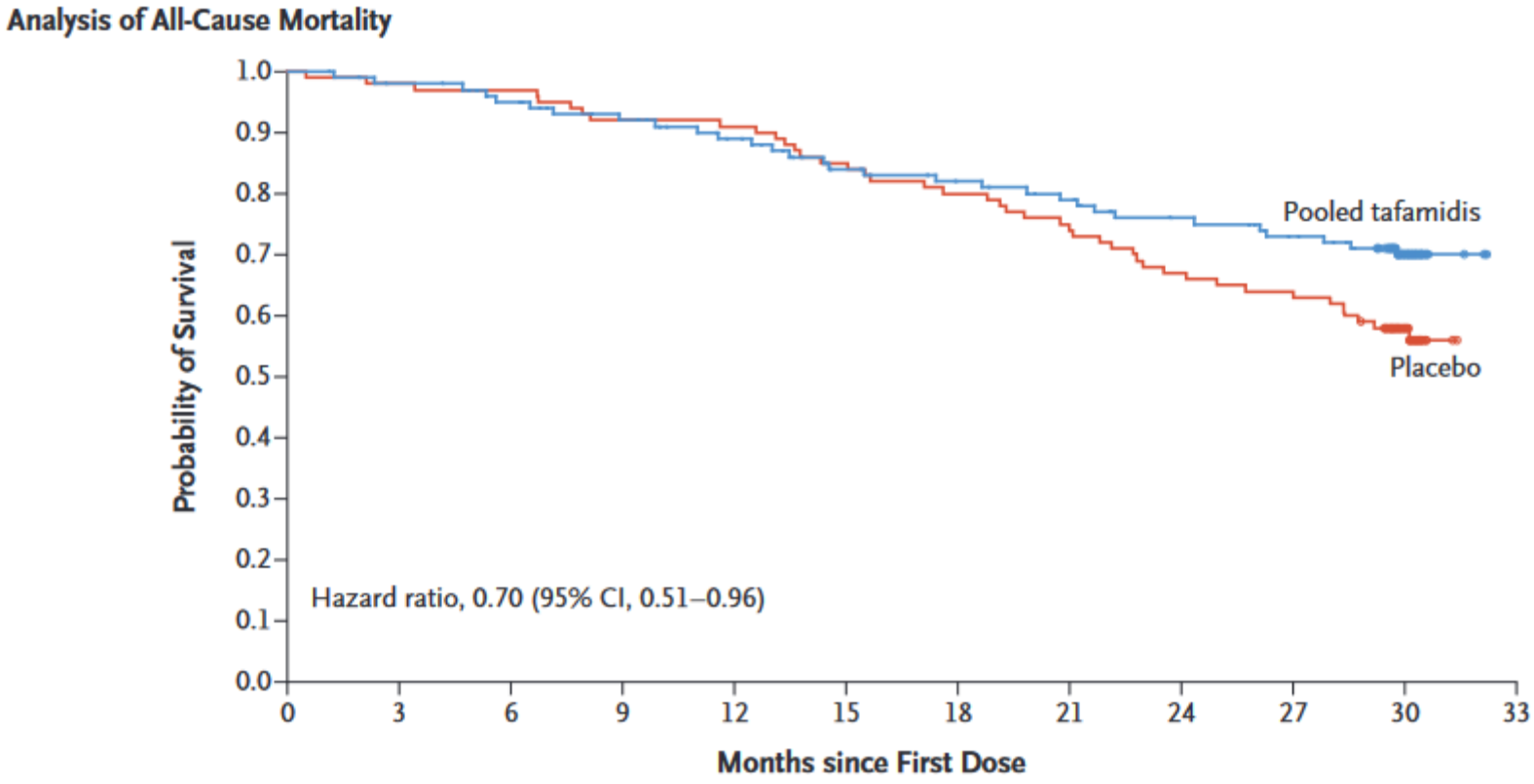
Tafamidis: ATTR-ACT



N = 441, mittleres Alter 75 Jahre

Maurer M, et al . N Engl J Med 2018;379:1007-16 (7)

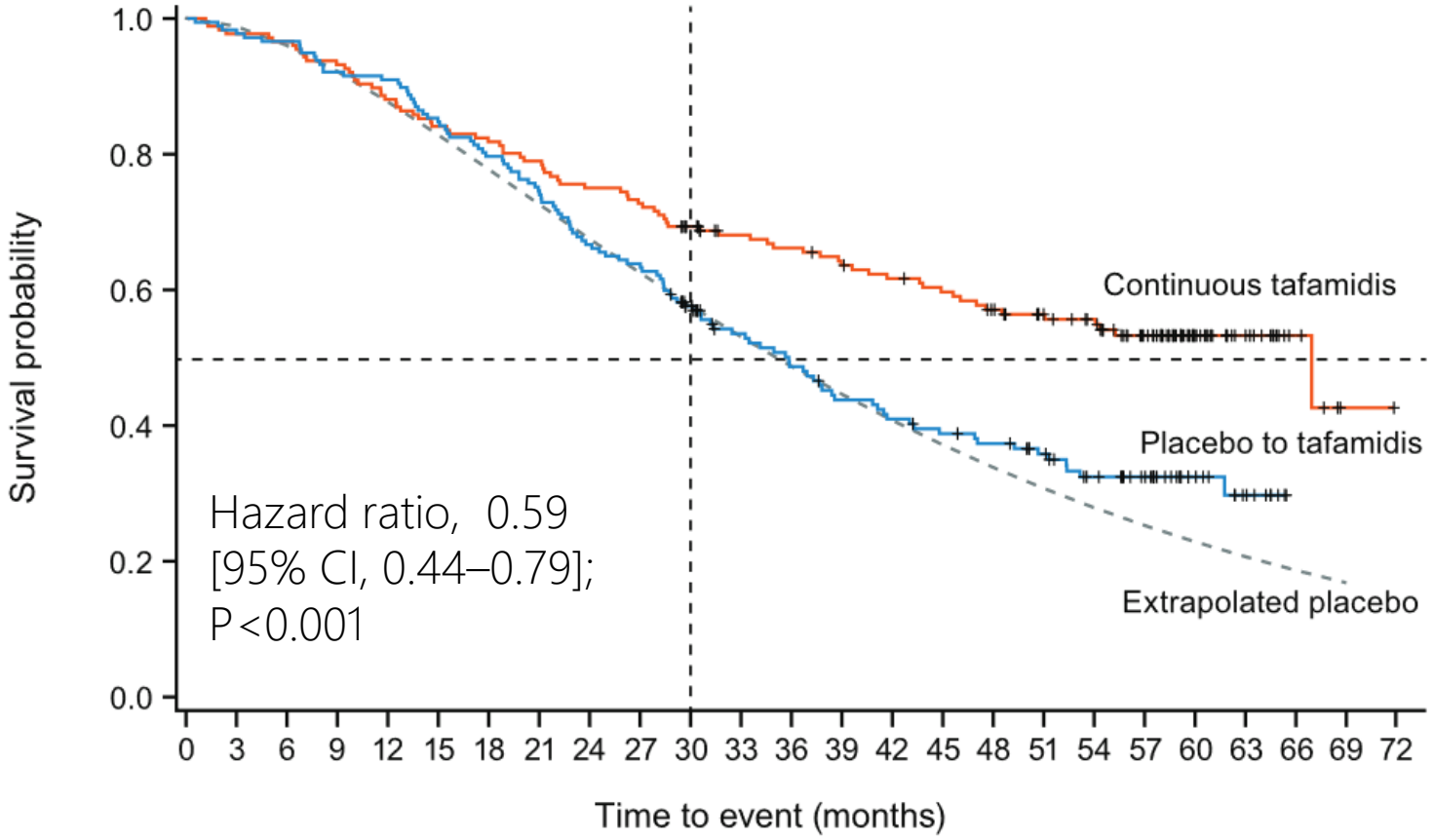
Tafamidis: ATTR-ACT



N = 441, mittleres Alter 75 Jahre

Maurer M, et al . N Engl J Med 2018;379:1007-16 (7)

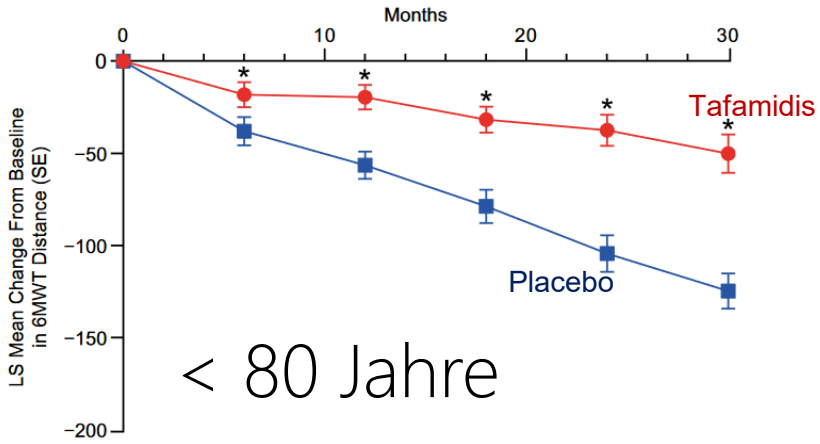
Tafamidis: ATTR-ACT 5 Jahre

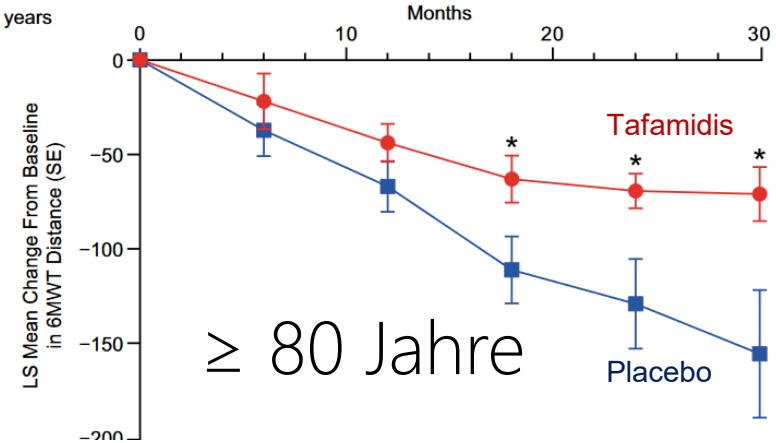


N = 353, mittleres Alter 75 Jahre

Elliott P, et al. Circ Heart Fail. 2022;15:e008193 (8)

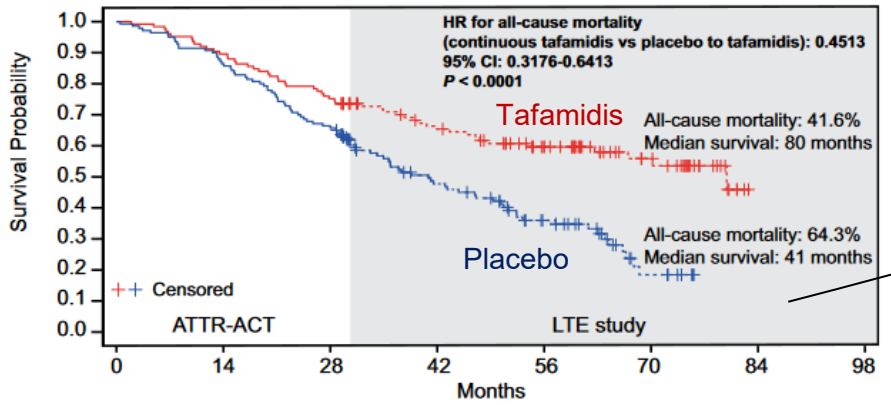
Tafamidis: Octogenarians





Garcia-Pavia P, et al. J Am Coll Cardiol HF 2024;12:150–160 (9)

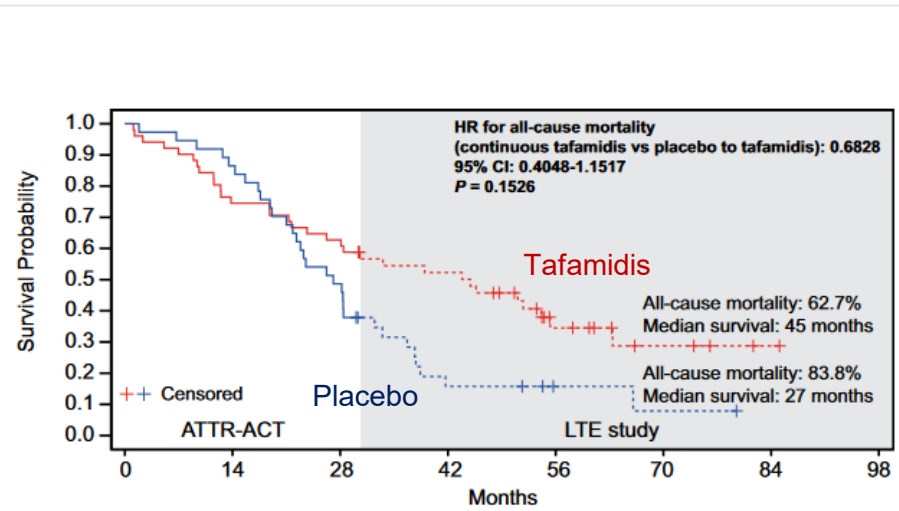
Tafamidis: Octogenarians



Mittleres Überleben 80 vs 41 Monate;

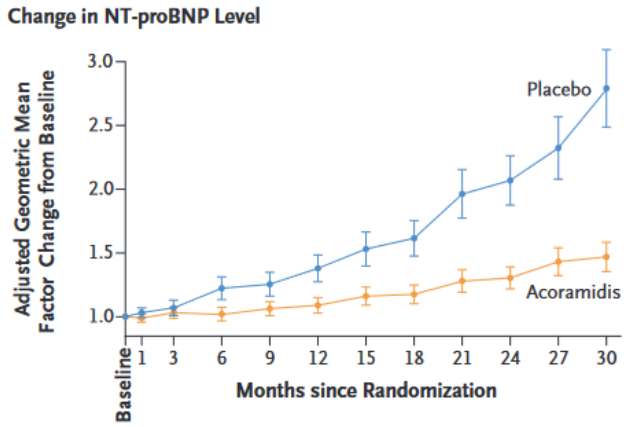
HR 0.4513

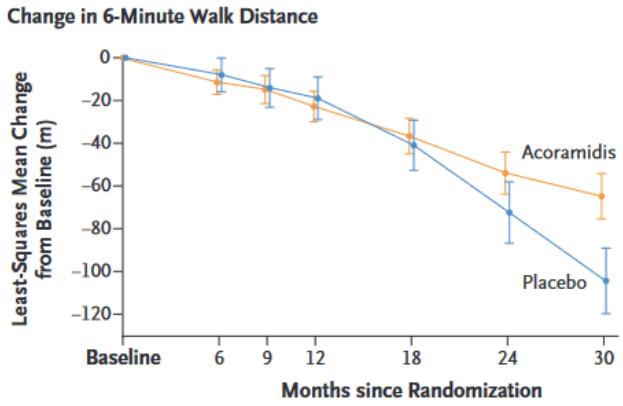
[95% CI: 0.3176-0.6413]; P < 0.0001)

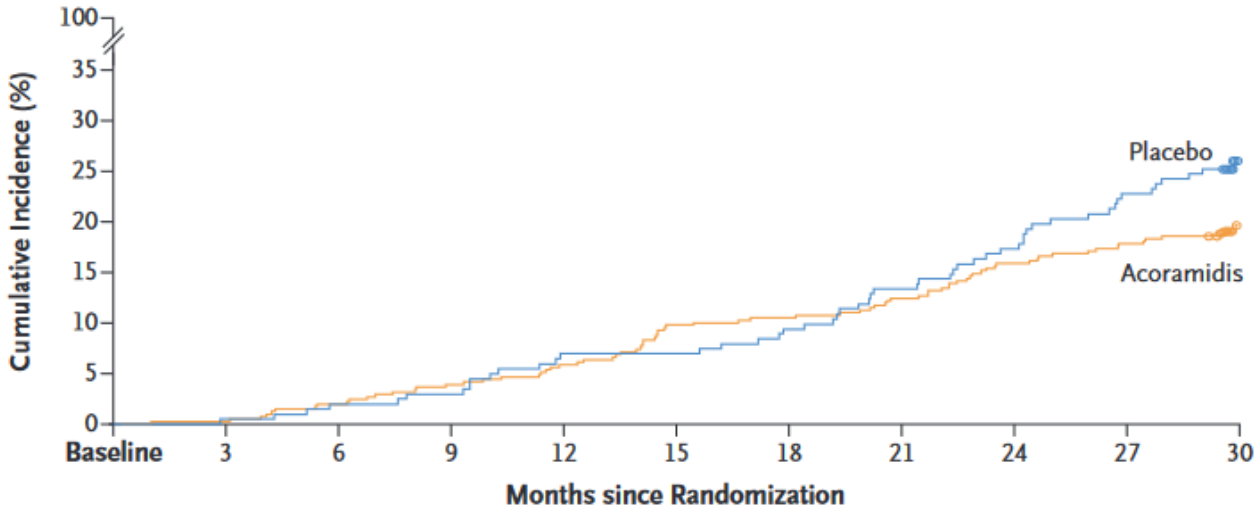


Garcia-Pavia P, et al. J Am Coll Cardiol HF 2024;12:150–160 (9)

Acoramidis: ATTRibute-CM



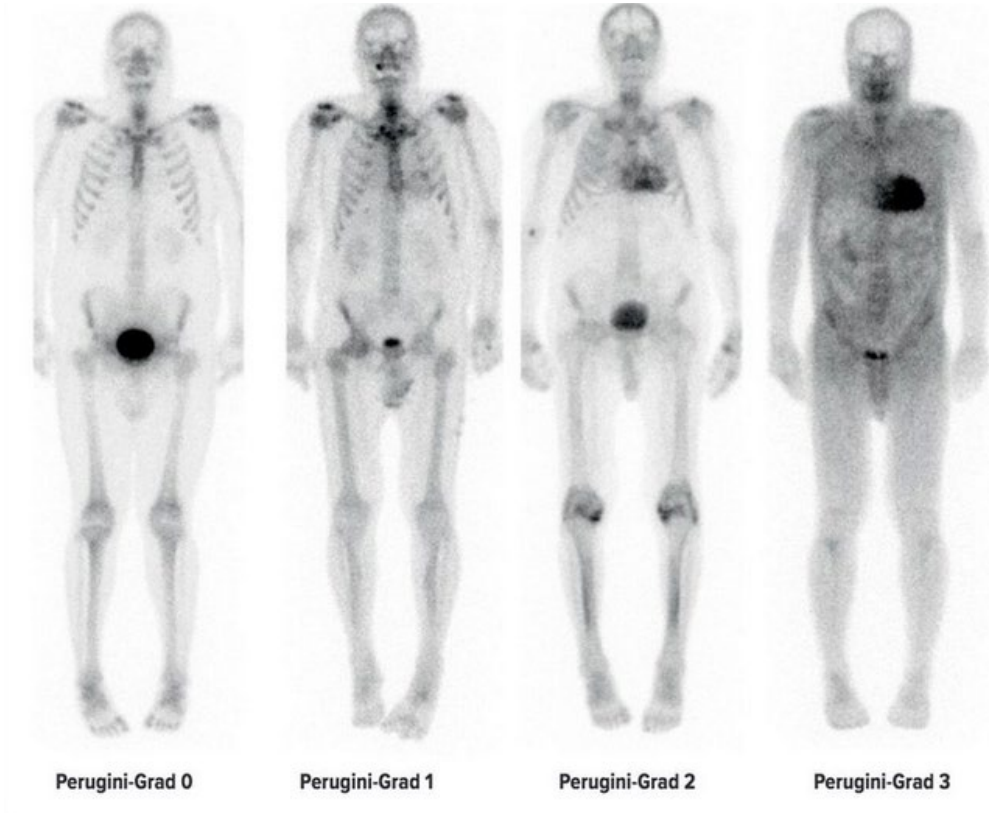




Gillmore J, et al. N Engl J Med 2024;390:132-42 (10)

Therapie mit Tafamidis

TTR-Amyloidose (Szintigraphie Perugini Score 2/3)



Therapie mit Tafamidis

TTR-Amyloidose (Szintigraphie Perugini Score 2/3)

NYHA I und II (NYHA III im Ermessen der Ärztin/des Arztes)

Monatliches Rezept

Empfehlung: „von einem in der Behandlung erfahrenen Arzt“

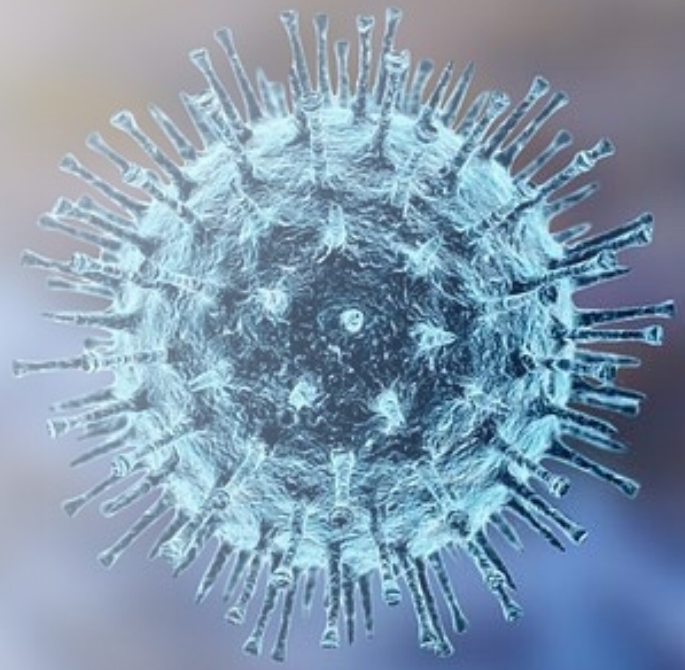
Myokarditis

Myokarditis: 12 Monate nach COVID

Meta Analyse

1.245 Millionen
nach COVID 19

19.630 Millionen
kein COVID-19



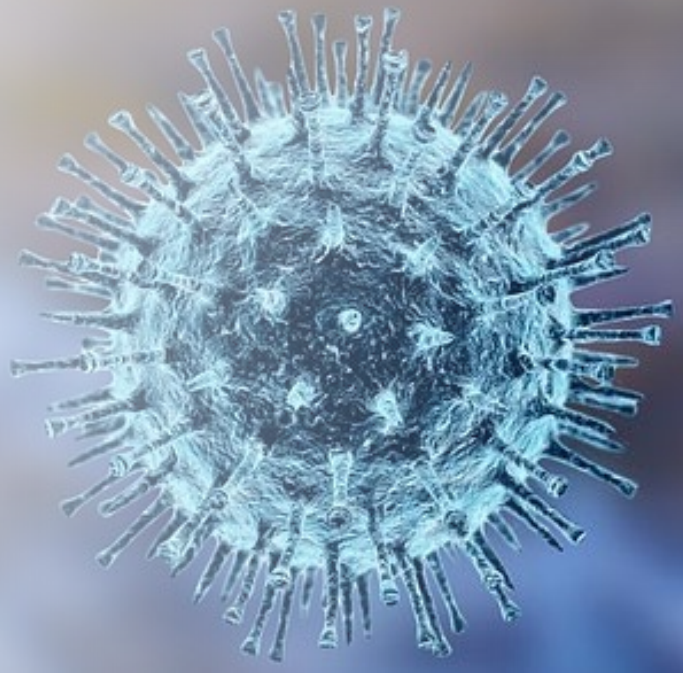
Zuin M, et al. Can J Cardiol. 2023;39(6):839-844

Myokarditis: 12 Monate nach COVID

Meta Analyse

1.245 Millionen
nach COVID 19

19.630 Millionen
kein COVID-19



MYOKARDITIS
0.21/1000 vs.
0.09/1000
HR 5.16 (3.9-6.9)

Zuin M, et al. Can J Cardiol. 2023;39(6):839-844

Myokarditis

Rate of cardiovascular events up to 8 years after uncomplicated myocarditis

A nationwide population based cohort study

Long-term outcomes after discharge

1,439 patients who had a first-time hospitalization with uncomplicated acute myocarditis and no known heart disease

were compared to

1,439 surgical controls after appendectomy

after propensity-score matching

Cardiovascular composite outcome

A composite of rehospitalization for myocarditis, pericardial disease, heart failure and its complications, cardiac arrhythmia, implantation of cardiac devices and heart transplant.

Schelldorfer A, et a. Eur Heart J Acute Cardiovasc Care. 2024 Feb 14:zuae021 (12)

Myokarditis

Rate of cardiovascular events up to 8 years after uncomplicated myocarditis

A nationwide population based cohort study

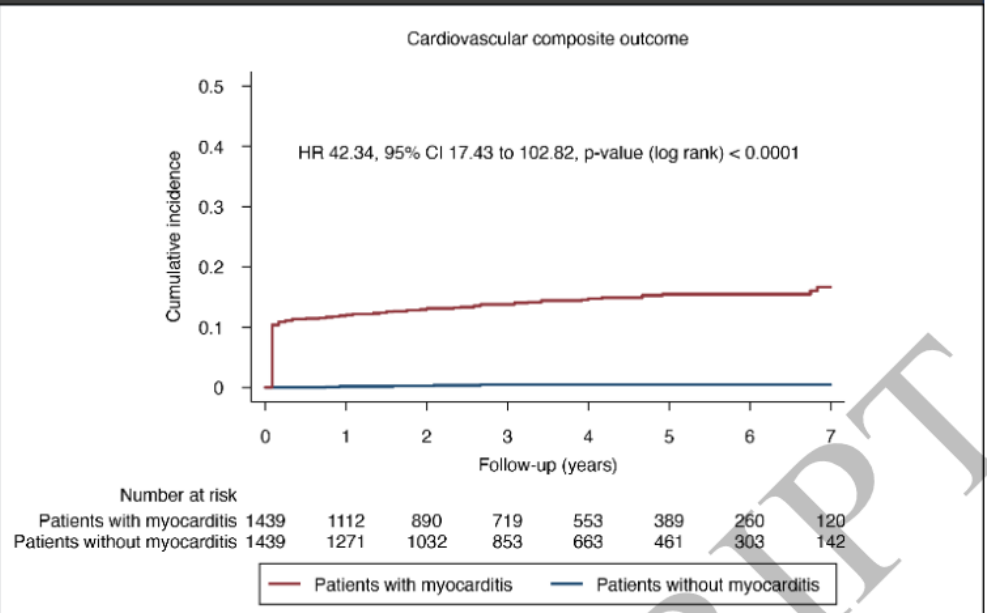
Long-term outcomes after discharge

1,439 patients who had a first-time hospitalization with uncomplicated acute myocarditis and no known heart disease

were compared to

1,439 surgical controls after appendectomy

after propensity-score matching



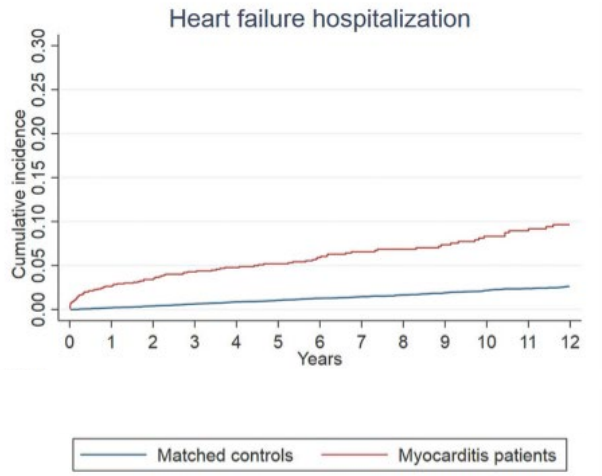
Cardiovascular composite outcome

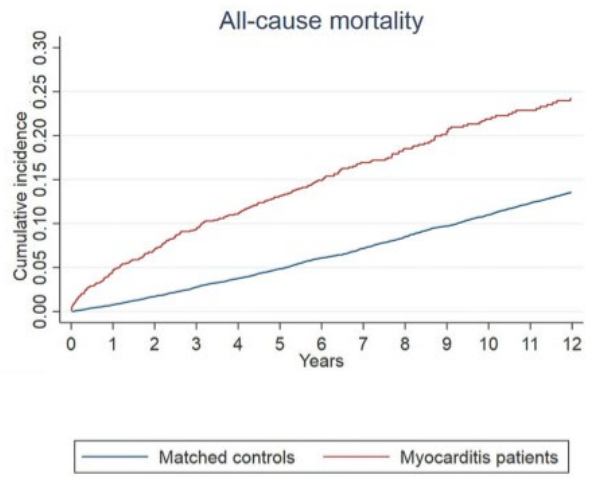
A composite of rehospitalization for myocarditis, pericardial disease, heart failure and its complications, cardiac arrhythmia, implantation of cardiac devices and heart transplant.

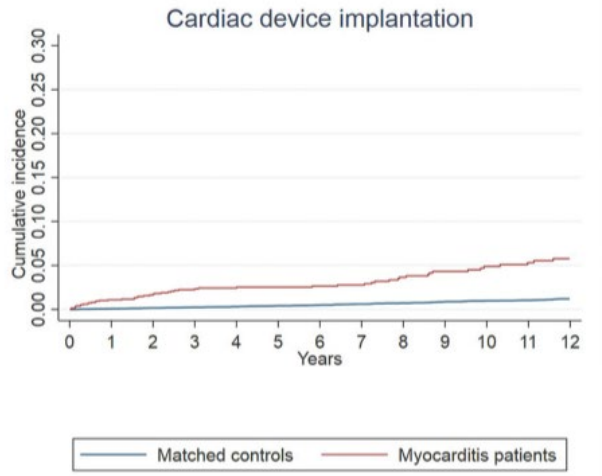
Patients with uncomplicated myocarditis	Patients without myocarditis (surgical controls)
Incidence rate per 1,000 person-years	
43.7	0.9
Hazard ratio 42.3 (95% confidence interval 17.4 to 102.8)	

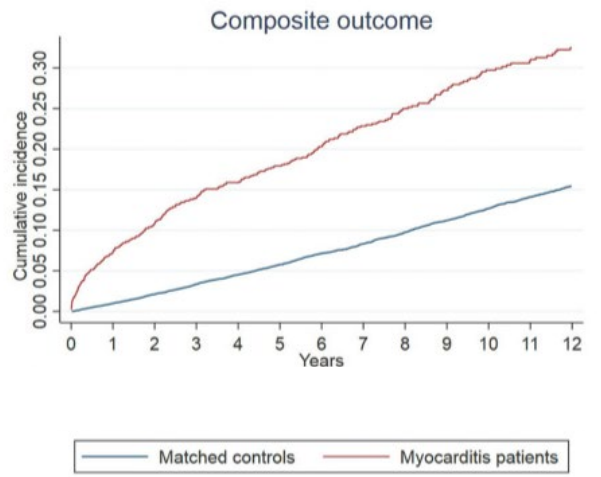
Schelldorfer A, et a. Eur Heart J Acute Cardiovasc Care. 2024 Feb 14:zuae021 (12)

Myokarditis: n = 1557 vs. 15430 Kontrollen (39J)



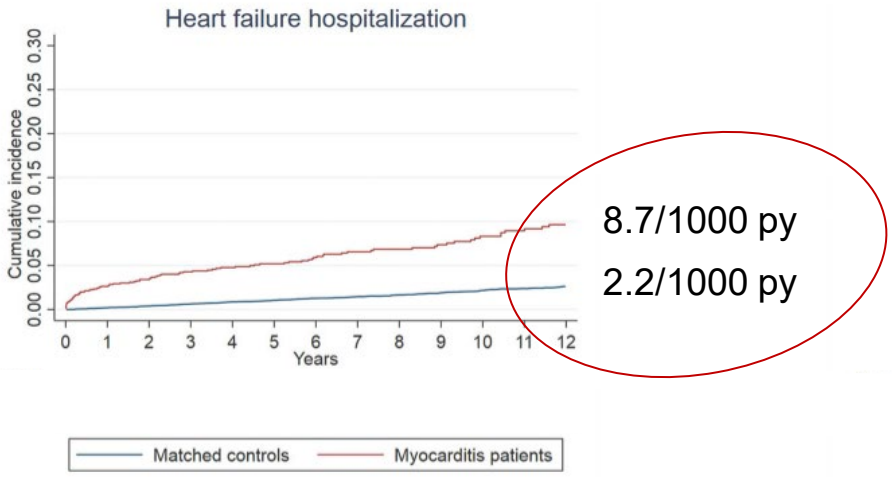






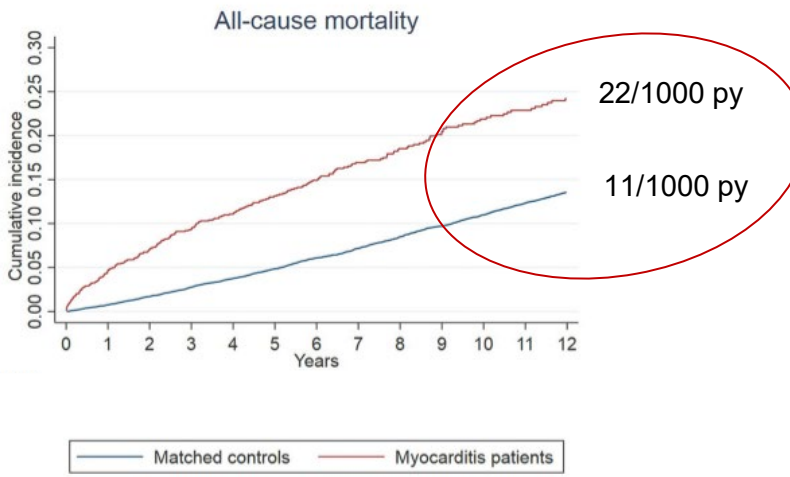
Ghanizada M, et al. Scand Cardiovasc J. 2021 Oct;55(5):264-269 (13)

Myokarditis: n = 1557 vs. 15430 Kontrollen (39J)



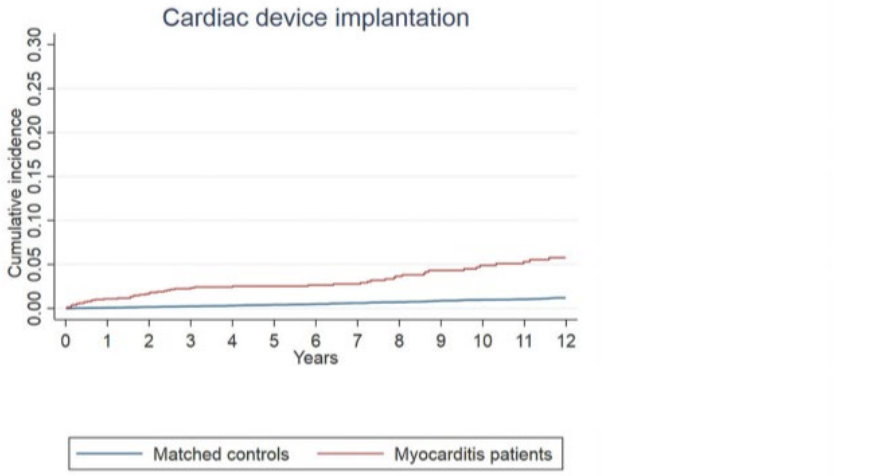
8.7/1000 py

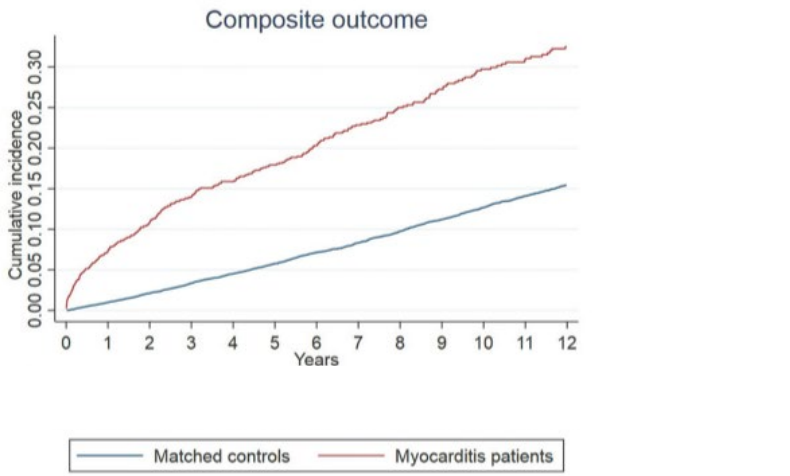
2.2/1000 py



22/1000 py

11/1000 py





Ghanizada M, et al. Scand Cardiovasc J. 2021 Oct;55(5):264-269 (13)

Literatur

1. Arbelo E, Protonotarios A, Gimeno JR, Arbustini E, Barriales-Villa R, Basso C, Bezzina CR, Biagini E, Blom NA, de Boer RA, De Winter T, Elliott PM, Flather M, Garcia-Pavia P, Haugaa KH, Ingles J, Jurcut RO, Klaassen S, Limongelli G, Loeys B, Mogensen J, Olivotto I, Pantazis A, Sharma S, Van Tintelen JP, Ware JS, Kaski JP; ESC Scientific Document Group. 2023 ESC Guidelines for the management of cardiomyopathies. Eur Heart J. 2023 Oct 1;44(37):3503-3626

2. Maron MS, Finley JJ, Bos JM, Hauser TH, Manning WJ, Haas TS, Lesser JR, Udelson JE, Ackerman MJ, Maron BJ. Prevalence, clinical significance, and natural history of left ventricular apical aneurysms in hypertrophic cardiomyopathy. Circulation. 2008 Oct 7;118(15):1541-9.

3. Desai MY, Owens A, Geske JB, Wolski K, Naidu SS, Smedira NG, Cremer PC, Schaff H, McErlean E, Sewell C, Li W, Sterling L, Lampl K, Edelberg JM, Sehnert AJ, Nissen SE. Myosin Inhibition in Patients With Obstructive Hypertrophic Cardiomyopathy Referred for Septal Reduction Therapy. J Am Coll Cardiol. 2022 Jul 12;80(2):95-108

4. Desai MY, Owens A, Wolski K, Geske JB, Saberi S, Wang A, Sherrid M, Cremer PC, Lakdawala NK, Tower-Rader A, Fermin D, Naidu SS, Smedira NG, Schaff H, McErlean E, Sewell C, Mudarris L, Gong Z, Lampl K, Sehnert AJ, Nissen SE. Mavacamten in Patients With Hypertrophic Cardiomyopathy Referred for Septal Reduction: Week 56 Results From the VALOR-HCM Randomized Clinical Trial. JAMA Cardiol. 2023 Oct 1;8(10):968-977

5. Maron MS, Masri A, Choudhury L, Olivotto I, Saberi S, Wang A, Garcia-Pavia P, Lakdawala NK, Nagueh SF, Rader F, Tower-Rader A, Turer AT, Coats C, Fifer MA, Owens A, Solomon SD, Watkins H, Barriales-Villa R, Kramer CM, Wong TC, Paige SL, Heitner SB, Kupfer S, Malik FI, Meng L, Wohltman A, Abraham T; REDWOOD-HCM Steering Committee and Investigators. Phase 2 Study of Aficamten in Patients With Obstructive Hypertrophic Cardiomyopathy. J Am Coll Cardiol. 2023 Jan 3;81(1):34-45

6. Grogan M, Wright RS. Decreasing Door-to-Diagnosis Time in Cardiac Amyloidosis: A Simple "One-Stop Shop" Approach. Mayo Clin Proc. 2023 Jan;98(1):7-10

7. Maurer MS, Schwartz JH, Gundapaneni B, Elliott PM, Merlini G, Waddington-Cruz M, Kristen AV, Grogan M, Witteles R, Damy T, Drachman BM, Shah SJ, Hanna M, Judge DP, Barsdorf AI, Huber P, Patterson TA, Riley S, Schumacher J, Stewart M, Sultan MB, Rapezzi C; ATTR-ACT Study Investigators. Tafamidis Treatment for Patients with Transthyretin Amyloid Cardiomyopathy. N Engl J Med. 2018 Sep 13;379(11):1007-1016

8. Elliott P, Drachman BM, Gottlieb SS, Hoffman JE, Hummel SL, Lenihan DJ, Ebede B, Gundapaneni B, Li B, Sultan MB, Shah SJ. Long-Term Survival With Tafamidis in Patients With Transthyretin Amyloid Cardiomyopathy. Circ Heart Fail. 2022 Jan;15(1):e008193.

9. Garcia-Pavia P, Sultan MB, Gundapaneni B, Sekijima Y, Perfetto F, Hanna M, Witteles R. Tafamidis Efficacy Among Octogenarian Patients in the Phase 3 ATTR-ACT and Ongoing Long-Term Extension Study. JACC Heart Fail. 2024 Jan;12(1):150-160

10. Gillmore JD, Judge DP, Cappelli F, Fontana M, Garcia-Pavia P, Gibbs S, Grogan M, Hanna M, Hoffman J, Masri A, Maurer MS, Nativi-Nicolau J, Obici L, Poulsen SH, Rockhold F, Shah KB, Soman P, Garg J, Chiswell K, Xu H, Cao X, Lystig T, Sinha U, Fox JC; ATTRibute-CM Investigators. Efficacy and Safety of Acoramidis in Transthyretin Amyloid Cardiomyopathy. N Engl J Med. 2024 Jan 11;390(2):132-142

11. Zuin M, Rigatelli G, Bilato C, Porcari A, Merlo M, Roncon L, Sinagra G. One-Year Risk of Myocarditis After COVID-19 Infection: A Systematic Review and Meta-analysis. Can J Cardiol. 2023 Jun;39(6):839-844

12. Schelldorfer A, Gregoriano C, Hauser S, Fuchs TA, Mueller B, Schuetz P, Kutz A. Rate of cardiovascular events up to 8 years after uncomplicated myocarditis: A nationwide cohort study. Eur Heart J Acute Cardiovasc Care. 2024 Feb 14:zuae021. doi: 10.1093/ehjacc/zuae021

13. Ghanizada M, Kristensen SL, Bundgaard H, Rossing K, Sigvardt F, Madelaire C, Gislason GH, Schou M, Hansen ML, Gustafsson F. Long-term prognosis following hospitalization for acute myocarditis - a matched nationwide cohort study. Scand Cardiovasc J. 2021 Oct;55(5):264-269

12. Schelldorfer A, Gregoriano C, Hauser S, Fuchs TA, Mueller B, Schuetz P, Kutz A. Rate of cardiovascular events up to 8 years after uncomplicated myocarditis: A nationwide cohort study. Eur Heart J Acute Cardiovasc Care. 2024 Feb 14:zuae021. doi: 10.1093/ehjacc/zuae021

13. Ghanizada M, Kristensen SL, Bundgaard H, Rossing K, Sigvardt F, Madelaire C, Gislason GH, Schou M, Hansen ML, Gustafsson F. Long-term prognosis following hospitalization for acute myocarditis - a matched nationwide cohort study. Scand Cardiovasc J. 2021 Oct;55(5):264-269