Praktische implementatie van cardiomyopathie richtlijn: regionaal DCM zorgpad

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Disclosures

Voor presentatie mogelijk relevante relaties	
Sponsoring of onderzoeksgeld:	geen
Honorarium of andere (financiële) vergoeding:	geen
Aandeelhouder:	geen
Andere relatie, namelijk …	geen

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

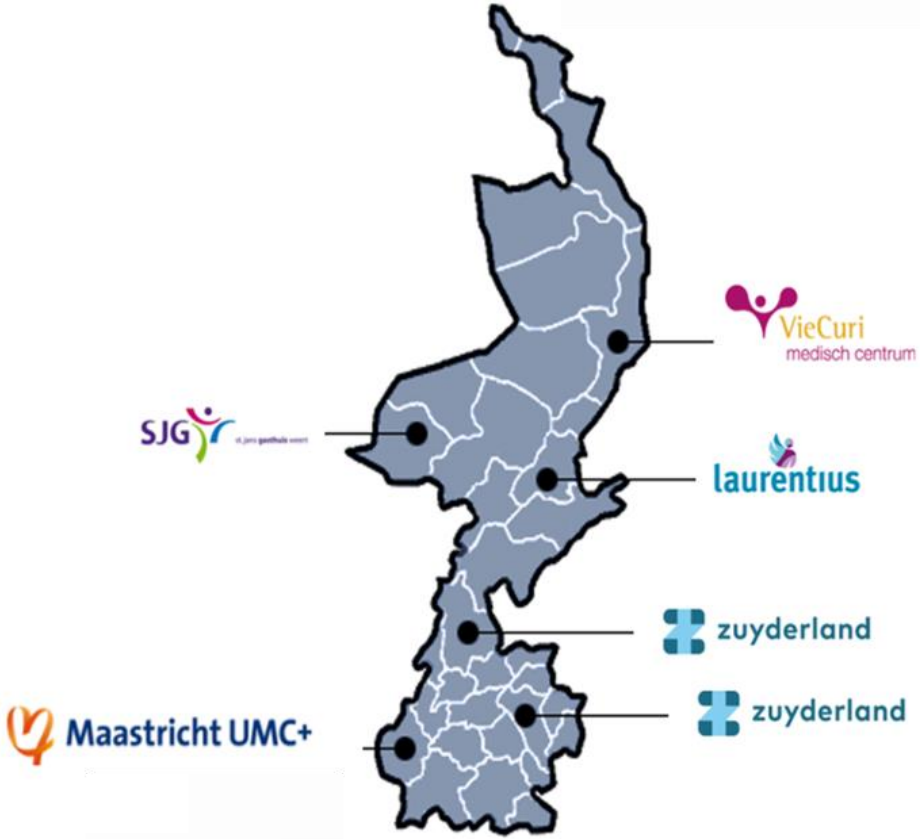
ESC GUIDELINES

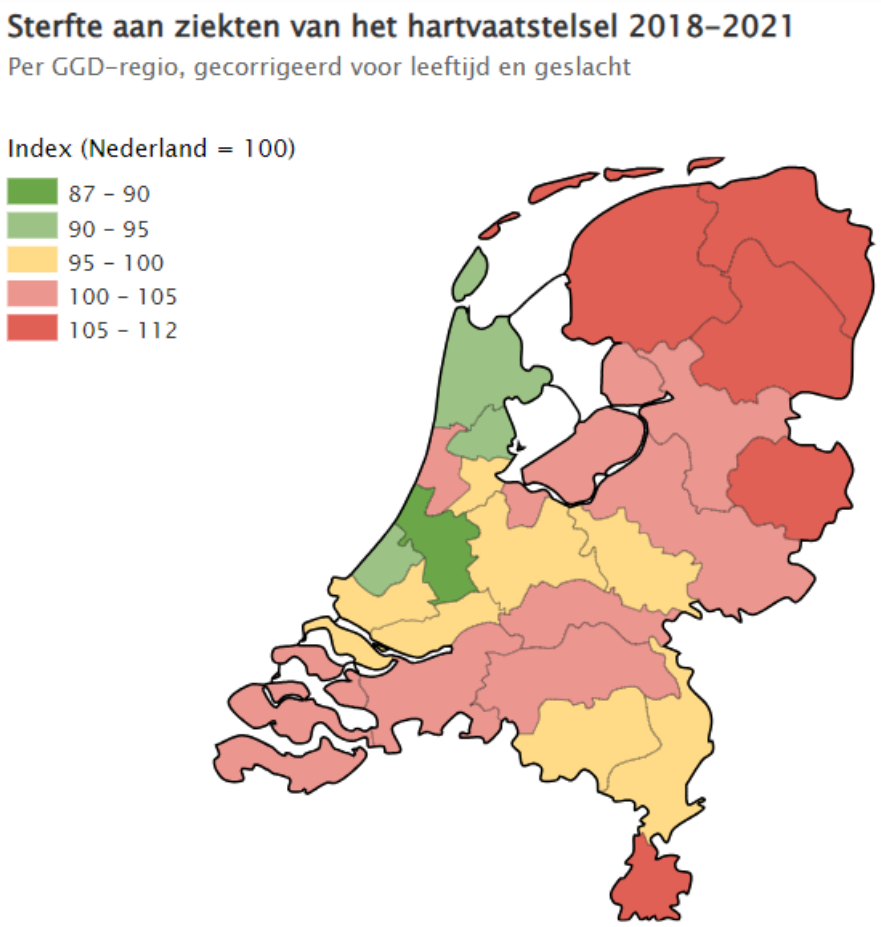
2023 ESC Guidelines for the management of cardiomyopathies

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

Sterke samenwerking Zuyderland – MUMC+





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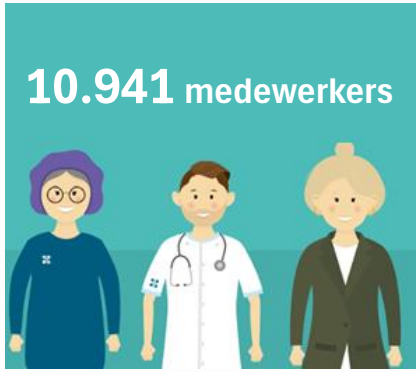
Zuyderland in notendop

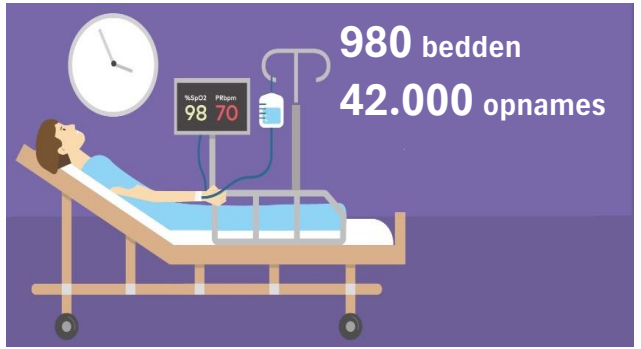
- Fusie 2015 Atrium (Heerlen) en Orbis MC (Sittard)

1. Sittard-Geleen
2. Heerlen
3. Kerkrade
4. Brunssum
5. Buitenpoli de Egthe



- 26 cardiologen: 7 HF
- HF combi-poli
- Cardio-onco poli
- HFpEF / PHT poli

- DCM poli
- 500-600 acute HF opn./j
- 800-850 nieuwe HF op combi poli /jaar



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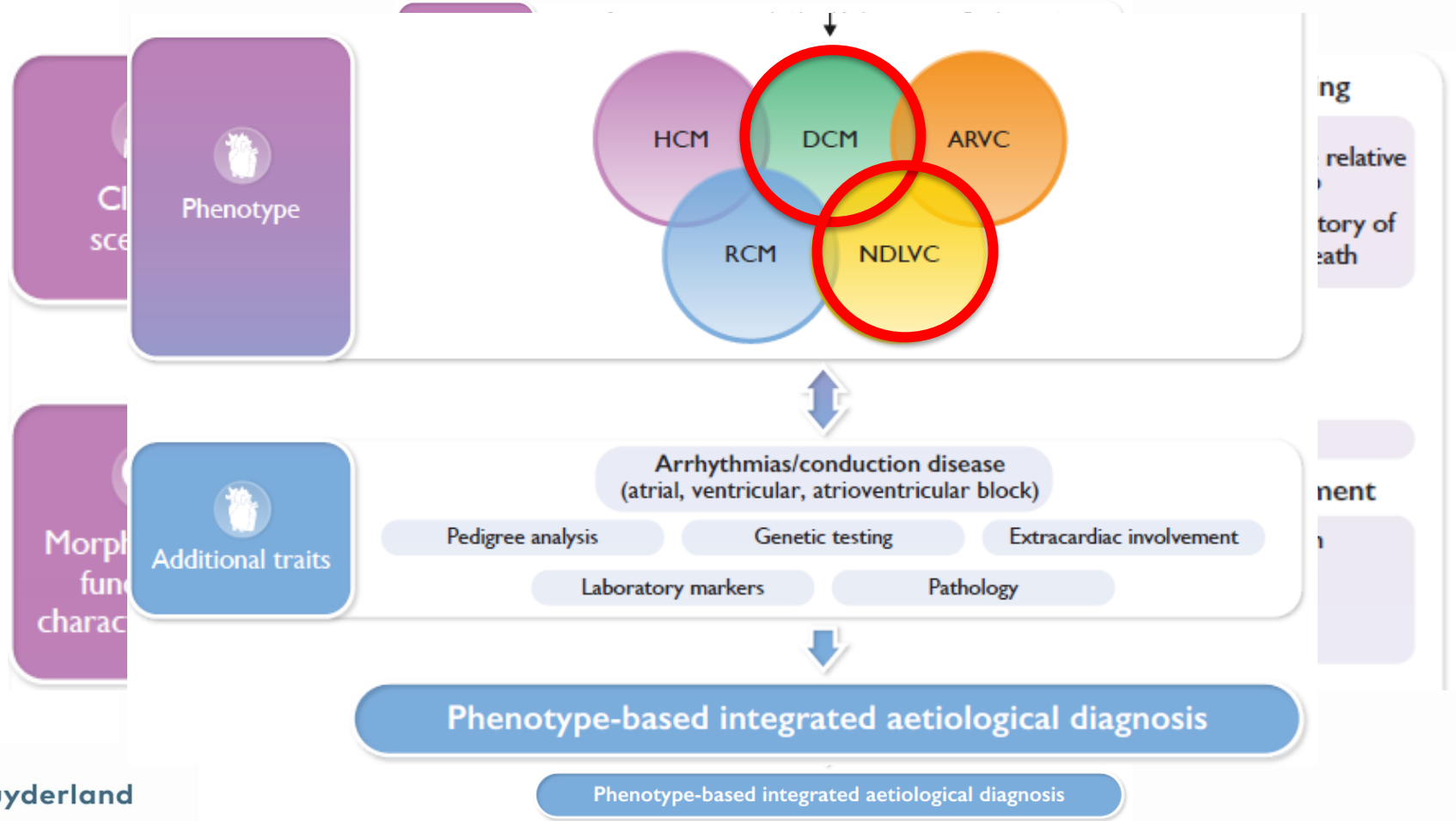
Regionaal DCM zorgpad



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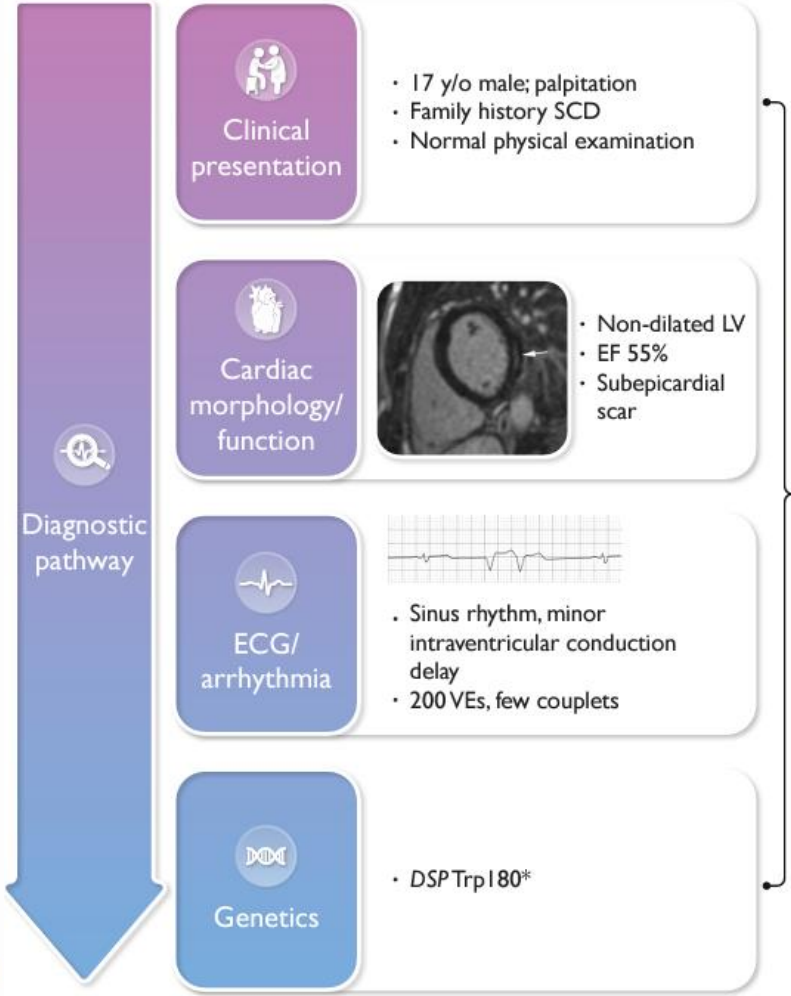

Recommendations	Class[a]	Level[b]
Contrast-enhanced CMR is recommended in patients with cardiomyopathy at initial evaluation.[10,90,116,119–143]	I	B
Contrast-enhanced CMR should be considered in patients with cardiomyopathy during follow-up to monitor disease progression and aid risk stratification and management.[89,90,120–122,127,129,136–147]	IIa	C
In families with cardiomyopathy in which a disease-causing variant has been identified, contrast-enhanced CMR should be considered in genotype-positive/phenotype-negative family members to aid diagnosis and detect early disease.[10,122,126,128,129,135–143,145,153–159]	IIa	B

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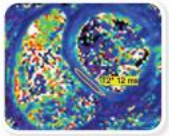
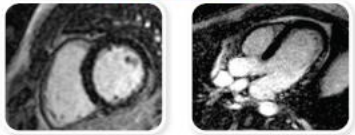
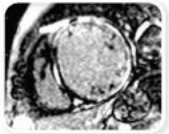
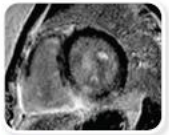
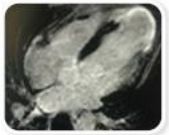
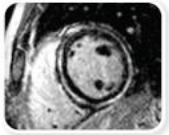
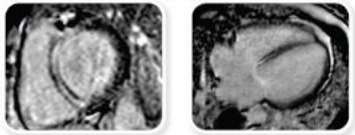
Cardiomyopathy phenotype	Finding	Cardiac CMR examples	Specific diseases to be considered
DCM	Short T2*		Haemochromatosis
	Subepicardial LGE		Post-myocarditis
	Lateral wall epicardial LGE		Dystrophinopathy
	Subepicardial and midwall LGE at basal septum +/- extension into inferolateral wall and RV insertion points		Sarcoidosis
	Apical transmural LGE		Chagas disease
NDLVC	Ring-like and/or subepicardial LGE pattern		*DSP* variants *FLNC* variants *DES* variants
	Septal mid-wall LGE		Laminopathy

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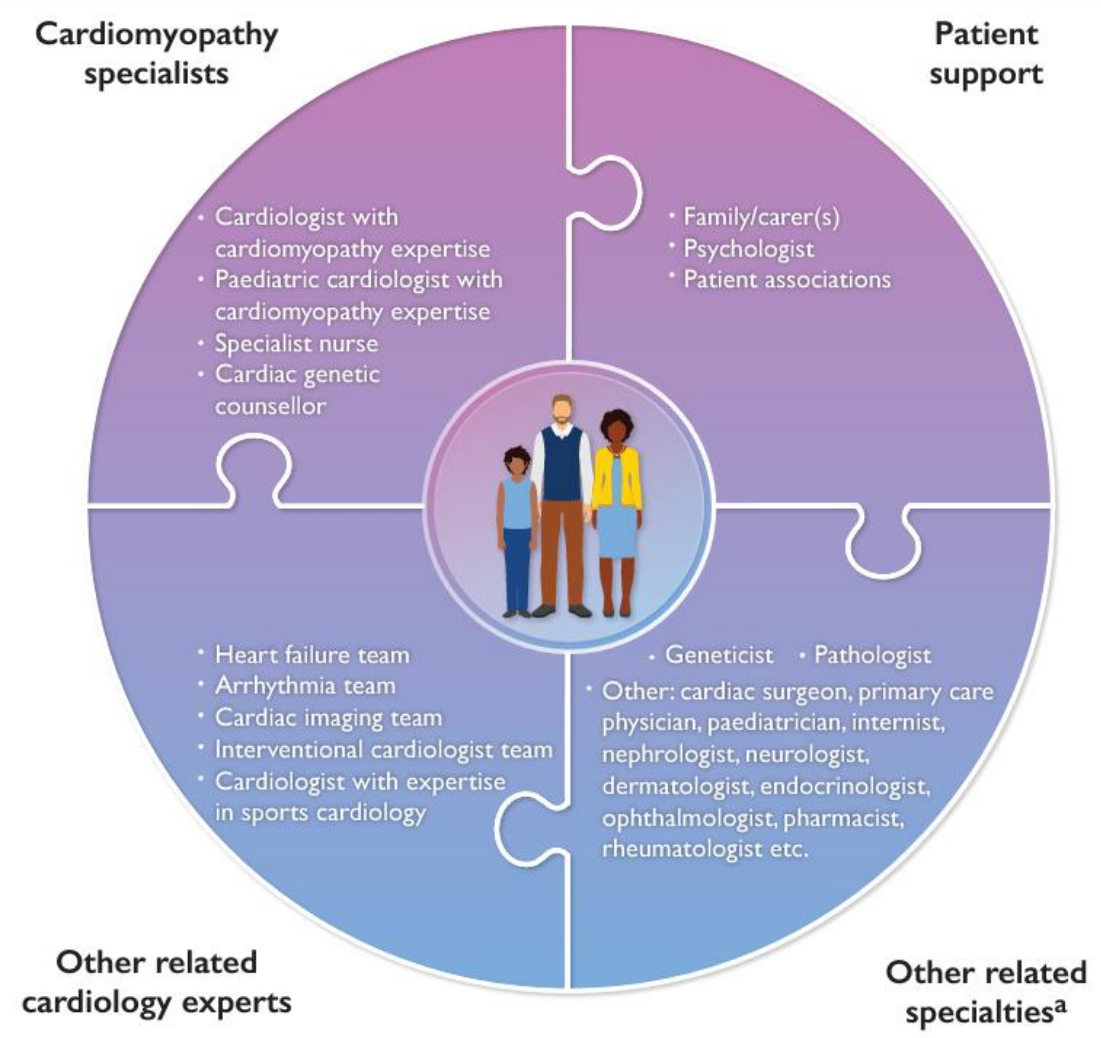
Cardiomyopathy specialists
Patient support
• Cardiologist with cardiomyopathy expertise
• Paediatric cardiologist with cardiomyopathy expertise
• Specialist nurse
• Cardiac genetic counsellor
• Family/carer(s)
• Psychologist
• Patient associations
• Heart failure team
• Arrhythmia team
• Cardiac imaging team
• Interventional cardiologist team
• Cardiologist with expertise in sports cardiology
• Geneticist • Pathologist
• Other: cardiac surgeon, primary care physician, paediatrician, internist, nephrologist, neurologist, dermatologist, endocrinologist, ophthalmologist, pharmacist, rheumatologist etc.
Other related cardiology experts
Other related specialties[a]

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Regionaal DCM zorgpad

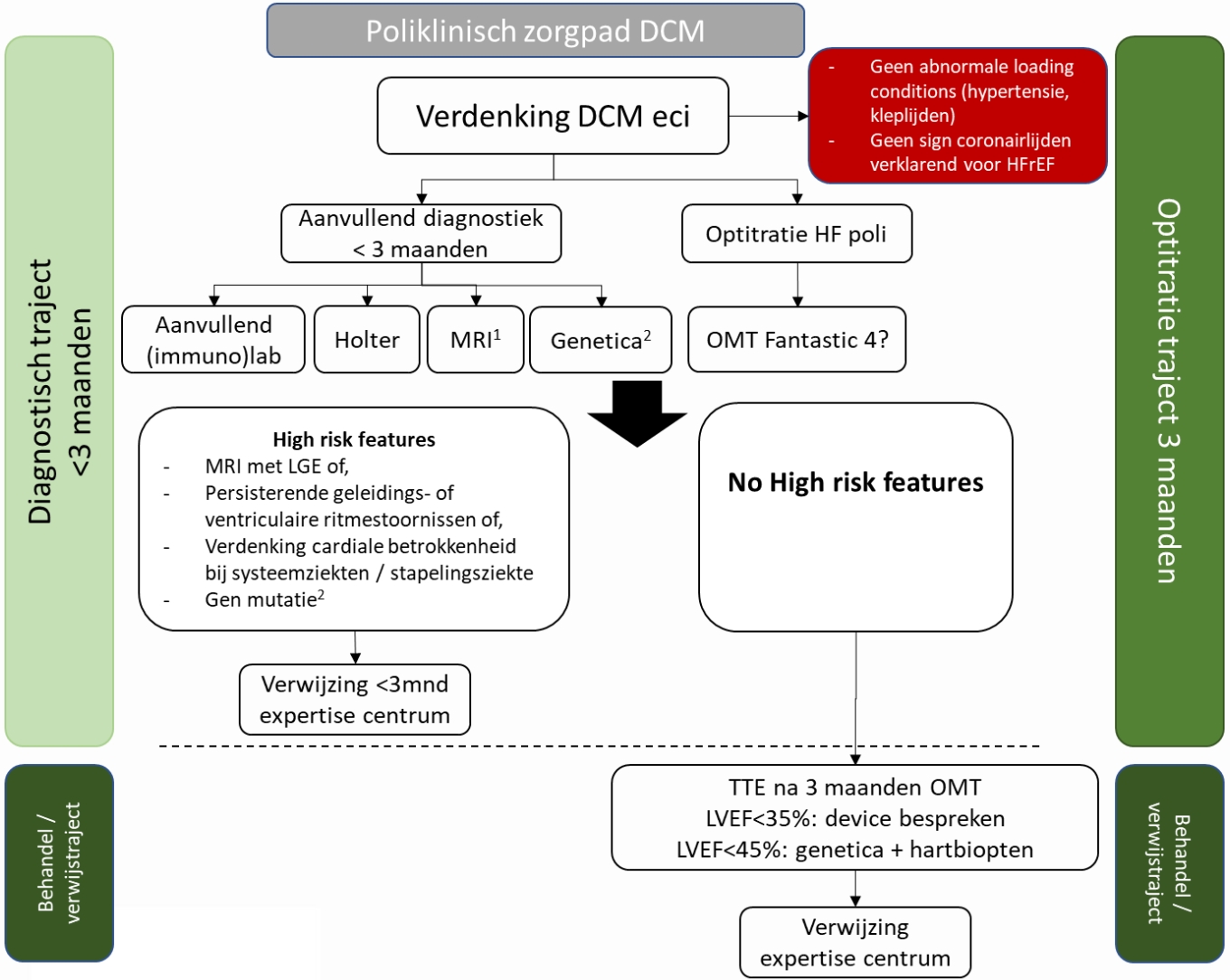
- Definities
  - **Gedilateerde cardiomyopathie (DCM)**
  - LV dilatatie en systolische dysfunctie die niet verklaard worden door
    - abnormale loading conditions (hypertensie, kleplijden) of coronarialijden, als voornaamste/enige oorzaak van hartfalen.
    - LVEF die lager is dan verwacht tov mate van ischemisch hartlijden (fibrose op MRI) of mate van kleplijden.

  - **Non-dilated left ventricular cardiomyopathie (NDLVC, 'precursor' van DCM)**
  - niet-ischemische LGE of fatty replacement zonder LV dilatatie met globale of regionale WBS (bijv post-myocarditis beeld)
  - geisoleerde globale LVEF<50% zonder LGE die niet verklaard worden door abnormale loading conditions (hypertensie, kleplijden) of coronarialijden

Doelen

- **Duidelijk omschreven zorgpad**
- **Effectieve communicatie tussen zorgverleners**
- **Welke onderzoeken, waar, wanneer en follow-up**
- **Vroege diagnostiek en behandeling DCM**
- **Onderwijs en training, wetenschappelijk onderzoek**

HOE ORGANISEREN WE DIT IN REGIO ZUID-LIMBURG?


Poliklinisch zorgpad DCM
Verdenking DCM eci
- Geen abnormale loading conditions (hypertensie, kleplijden)
- Geen sign coronairlijden verklarend voor HFrEF
Aanvullend diagnostiek < 3 maanden
Optitratie HF poli
Aanvullend (immuno)lab
Holter
MRI[1]
Genetica[2]
OMT Fantastic 4?
High risk features
- MRI met LGE of,
- Persisterende geleidings- of ventriculaire ritmestoornissen of,
- Verdenking cardiale betrokkenheid bij systeemziekten / stapelingsziekte
- Gen mutatie[2]
No High risk features
Verwijzing <3mnd expertise centrum
TTE na 3 maanden OMT
LVEF<35%: device bespreken
LVEF<45%: genetica + hartbiopten
Verwijzing expertise centrum
Diagnostisch traject <3 maanden
Optitratie traject 3 maanden
Behandel / verwijstraject
Behandel / verwijstraject

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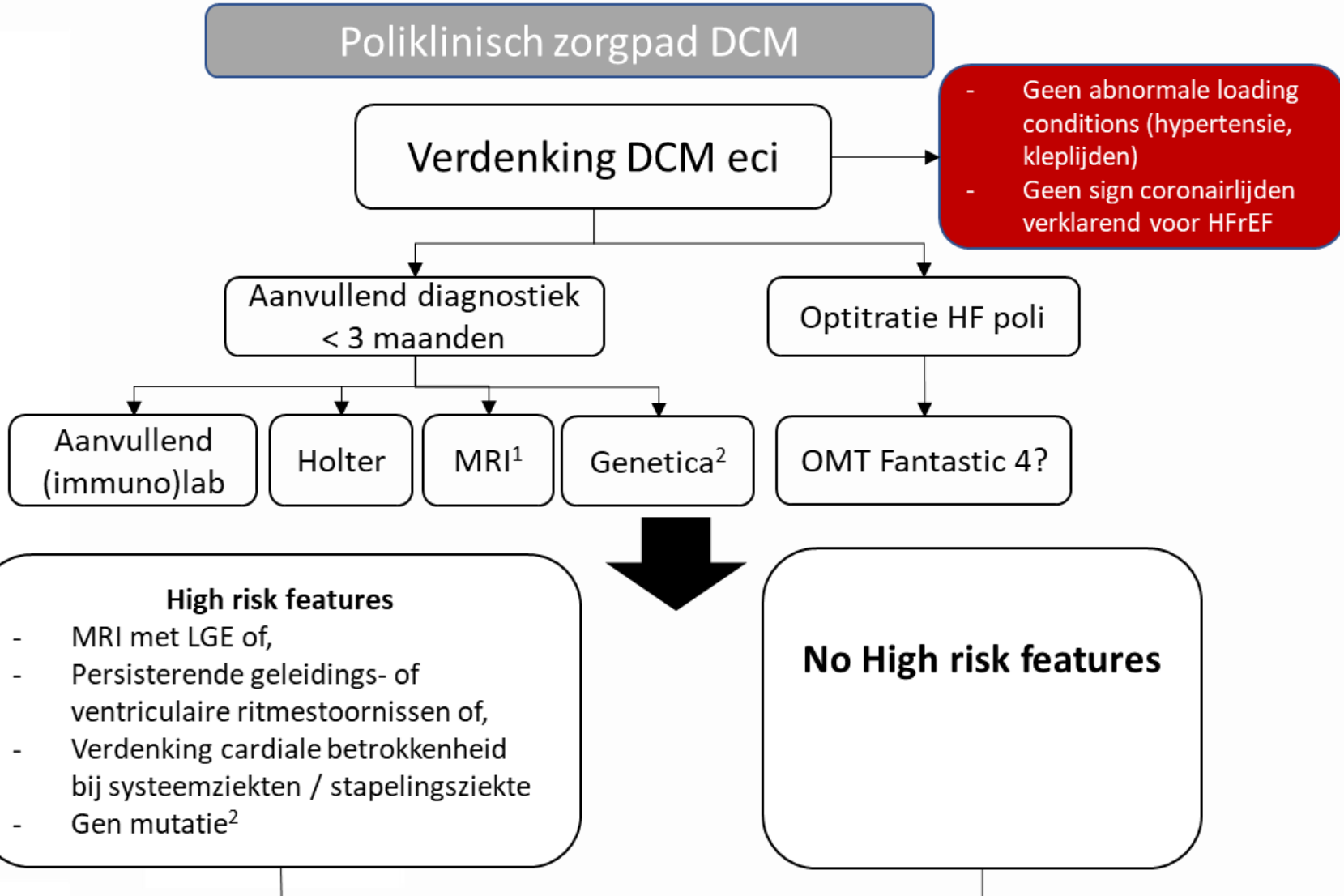
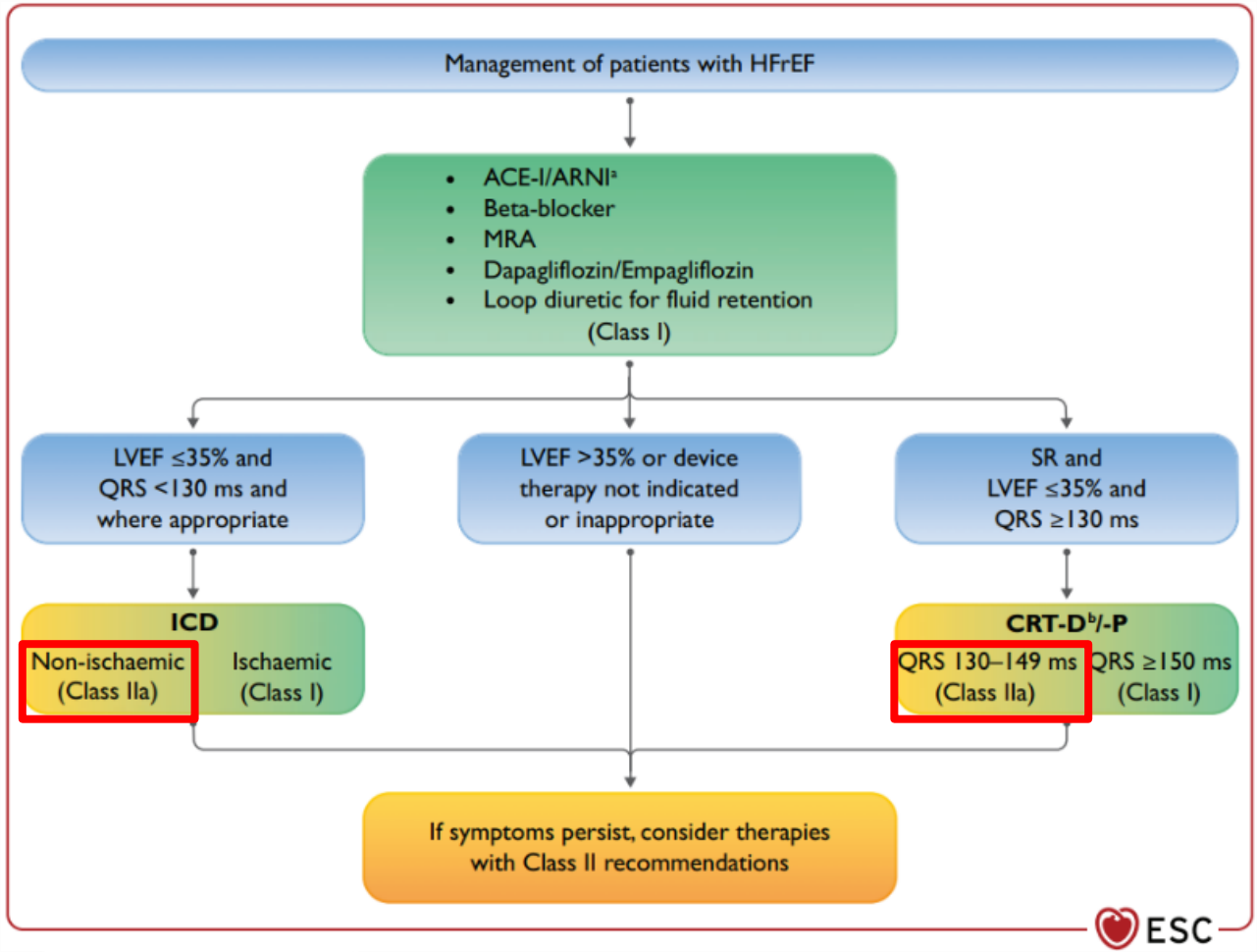
Poliklinisch zorgpad DCM



Device indicaties: ESC HF Guideline



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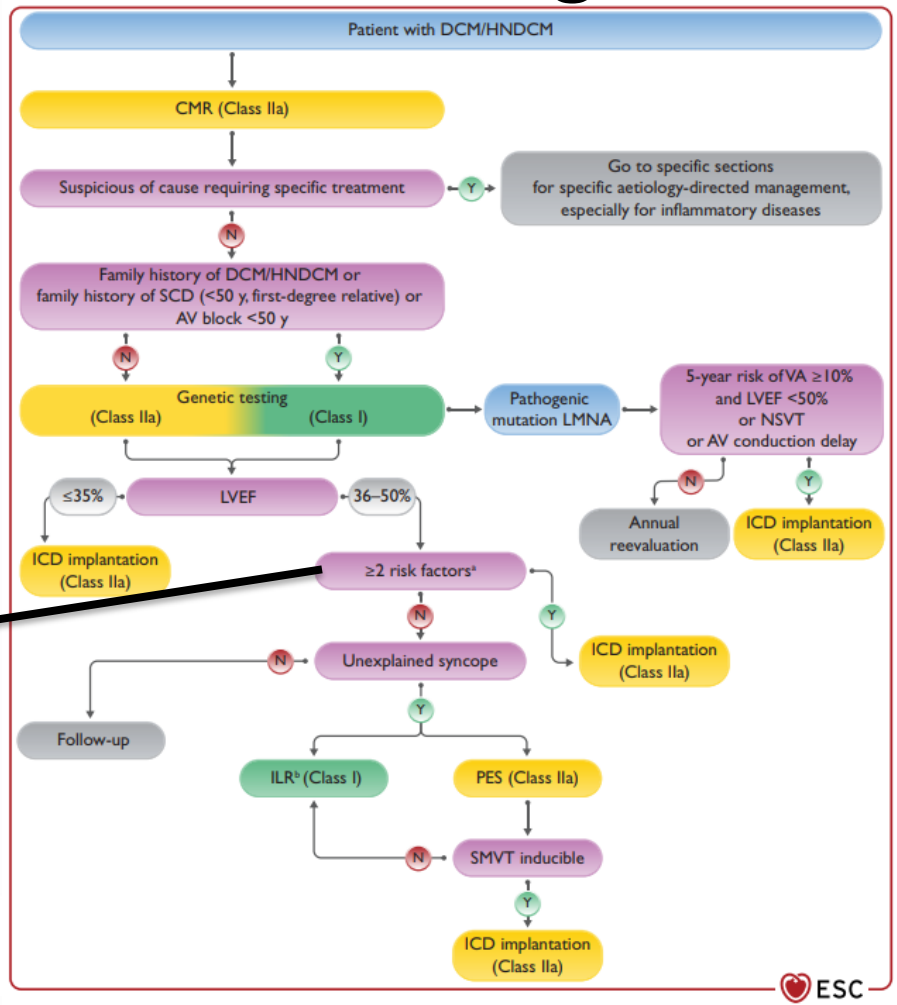
McDonagh et al.,EHJ 2021

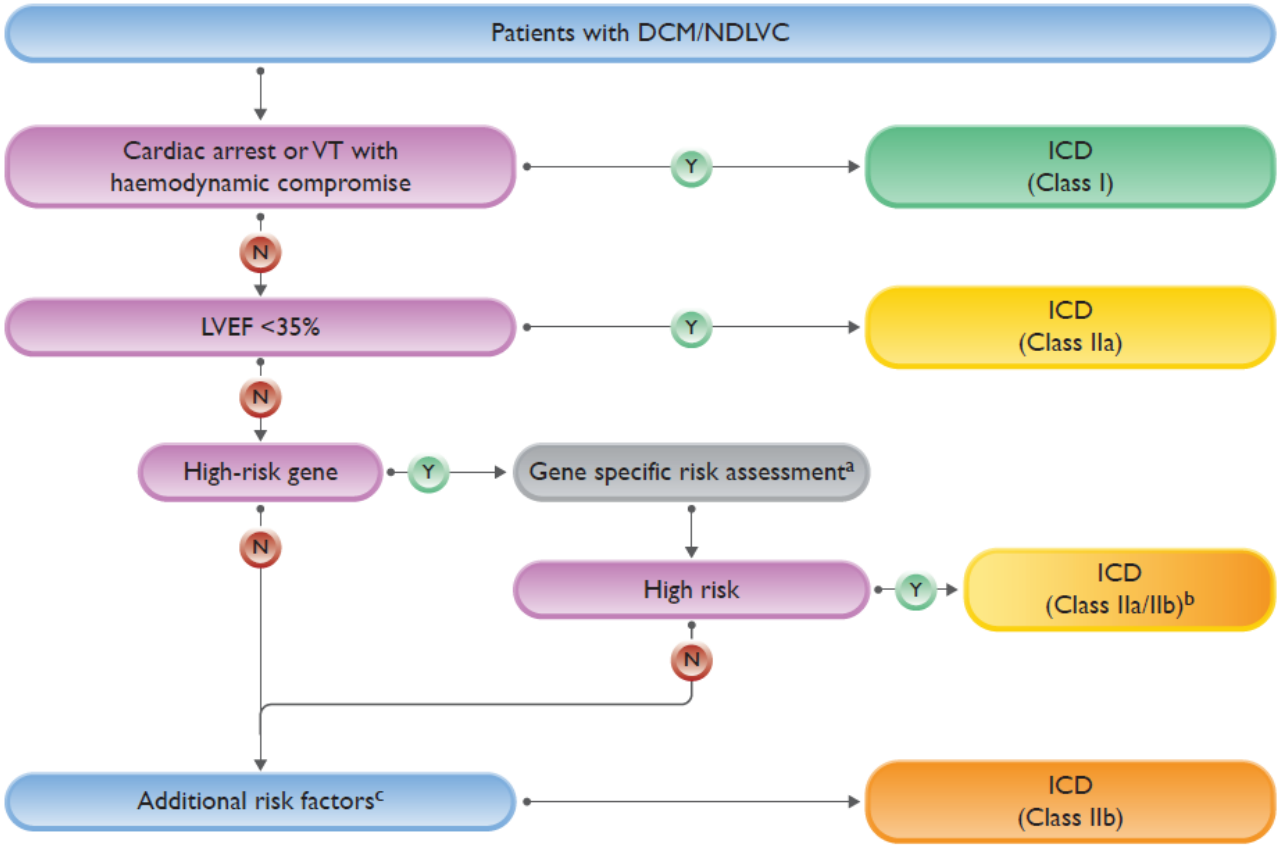
Device indicaties: changes in ESC VA/SCD

Risk factors:
- unexplained syncope
- pathogenic mut. PLN, FLNC, RBM20
- LGE on CMR
- inducible SMVT at PES



Zeppenfeld et al. EHJ 2022

Device indicaties: ESC CMP



High risk:

- LMNA
- FLNC
- PLN
- RBM20
- DSP
- TMEM43

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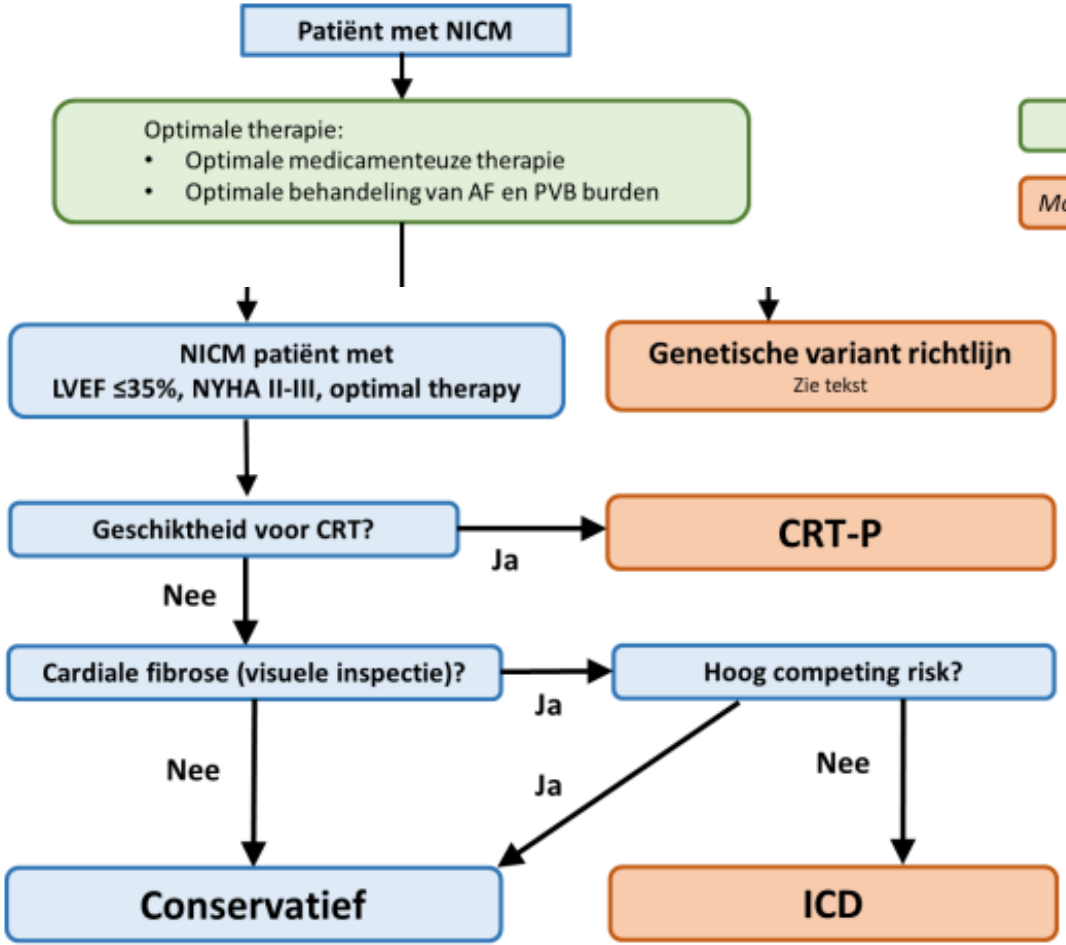
Arbelo et al. EHJ 2023

Device indicaties: NL



LMNA, PLN, FLNC, RBM20, DSP

http://www.hfmetascore.org
High competing risk:
- 1j overlijdensrisico >35%

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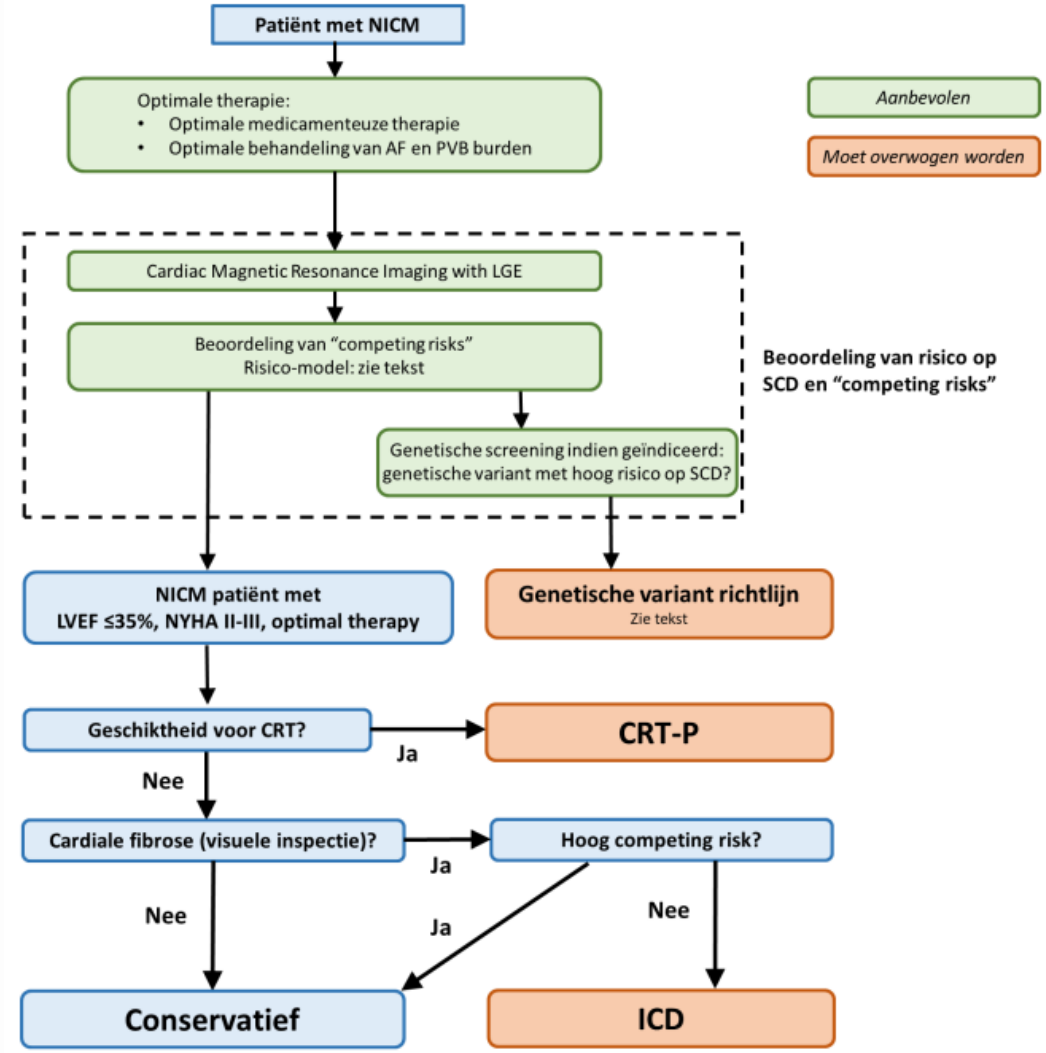
LVEF<35%, NYHA II-III zonder LGE?

(ESC 2022: class IIA)



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Hartbiopten

Recommendation	Class[a]	Level[b]
In patients with suspected cardiomyopathy, EMB should be considered to aid in diagnosis and management when the results of other clinical investigations suggest myocardial inflammation, infiltration, or storage that cannot be identified by other means.[174–177]	IIa	C

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Arbelo et al. EHJ 2023

Table 3 Indications for endomyocardial biopsy

Clinical presentation	Endomyocardial biopsy finding
• Suspected fulminant myocarditis or acute myocarditis with acute HF, LV dysfunction and/or rhythm disorders. • Suspected myocarditis in haemodynamically stable patients.	Myocarditis type: • Lymphocytic myocarditis • Eosinophilic myocarditis • Giant cell myocarditis • Granulomatous myocarditis
Dilated cardiomyopathy with recent onset HF, moderate-to-severe LV dysfunction, refractory to standard treatment (following exclusion of specific aetiologies).	Myocyte abnormalities, focal or diffuse fibrosis and inflammatory infiltrates (inflammatory cardiomyopathy).
Suspected ICI-mediated cardiotoxicity: acute HF with/without haemodynamic instability early after drug initiation (~ first 4 cycles)	ICI-mediated myocarditis
High-degree atrioventricular block, syncope and/or unexplained ventricular arrhythmias (ventricular fibrillation, ventricular tachycardia, frequent multifocal premature ventricular complexes), refractory to treatment, without obvious cardiac disease or with minimal structural abnormalities.	• Myocarditis • Arrhythmogenic right ventricular cardiomyopathy • Cardiac sarcoidosis
Autoimmune disorders with progressive HF unresponsive to treatment with/without sustained ventricular arrhythmias and/or conduction abnormalities.	• Autoimmune myocarditis • Viral myocarditis • Vasculitis/vasculopathy
MINOCA/takotsubo syndrome with progressive LV dysfunction and HF with/without ventricular arrhythmias or conduction abnormalities.	Differential diagnosis of myocarditis
Unexplained restrictive or hypertrophic cardiomyopathy.	• Amyloidosis • Infiltrative/storage disorders (Anderson–Fabry disease, glycogen storage diseases, sarcoidosis, haemochromatosis)
Cardiac tumours.	Histopathological diagnosis
• Routine surveillance EMB • Symptom-triggered EMB	HTx rejection status

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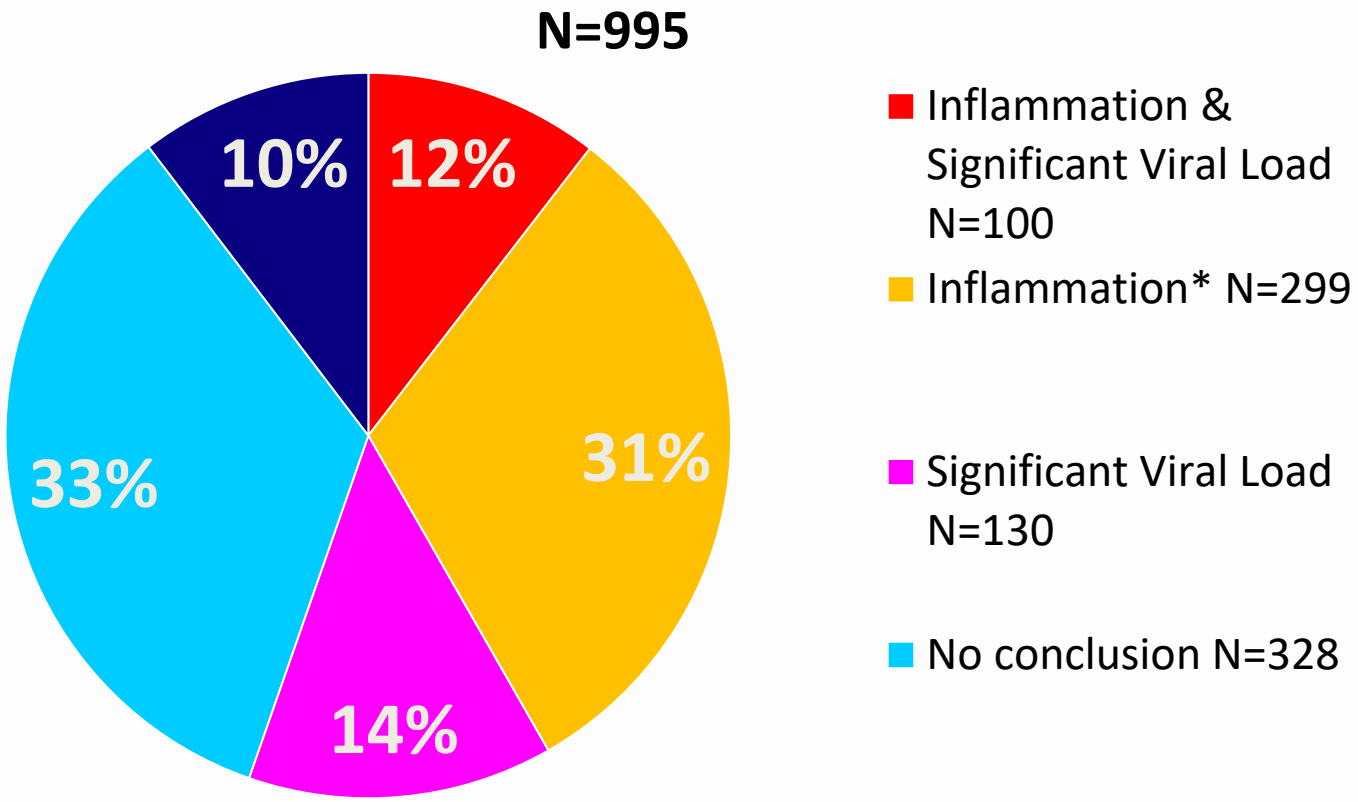
Seferovic et al. EJHF 2021

Diagnostic yield EMB



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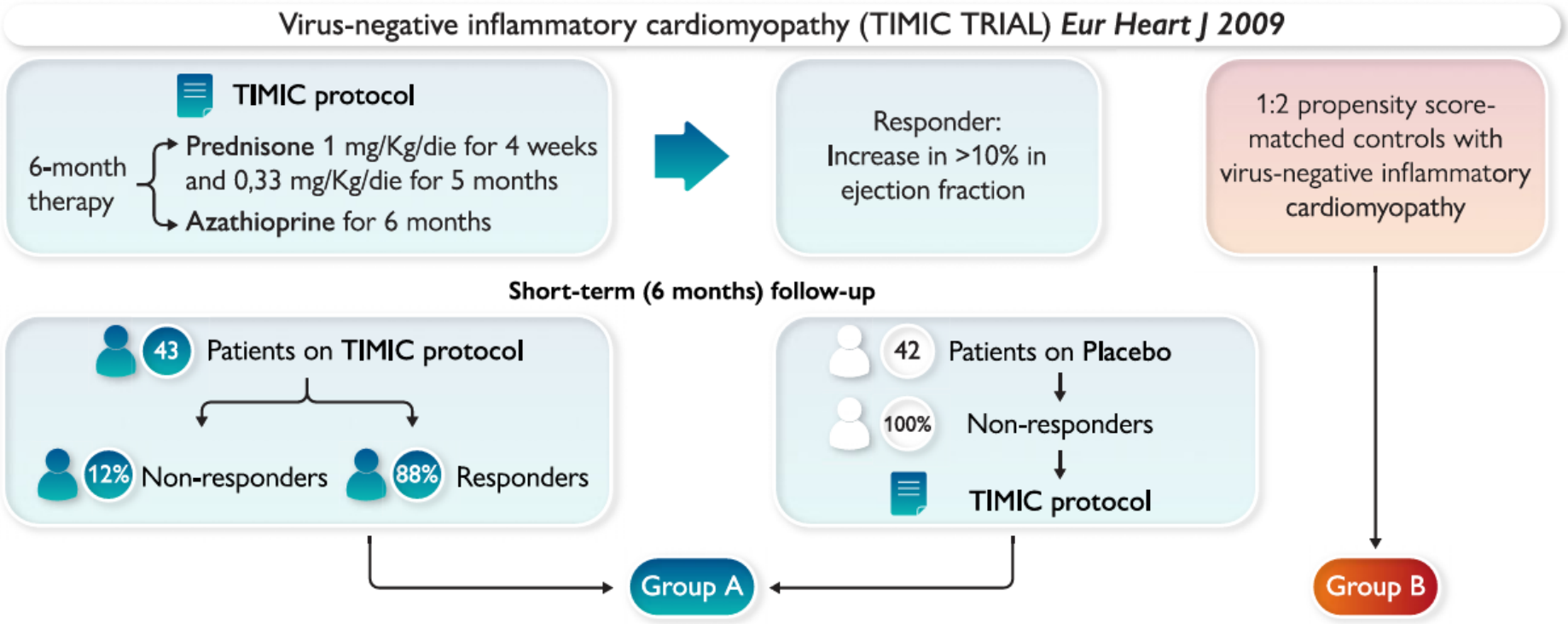
Hazebroek et al., JACC 2015. Henkens et al. ESC HF 2022

Immuunsuppressie bij inflammatoire CMP



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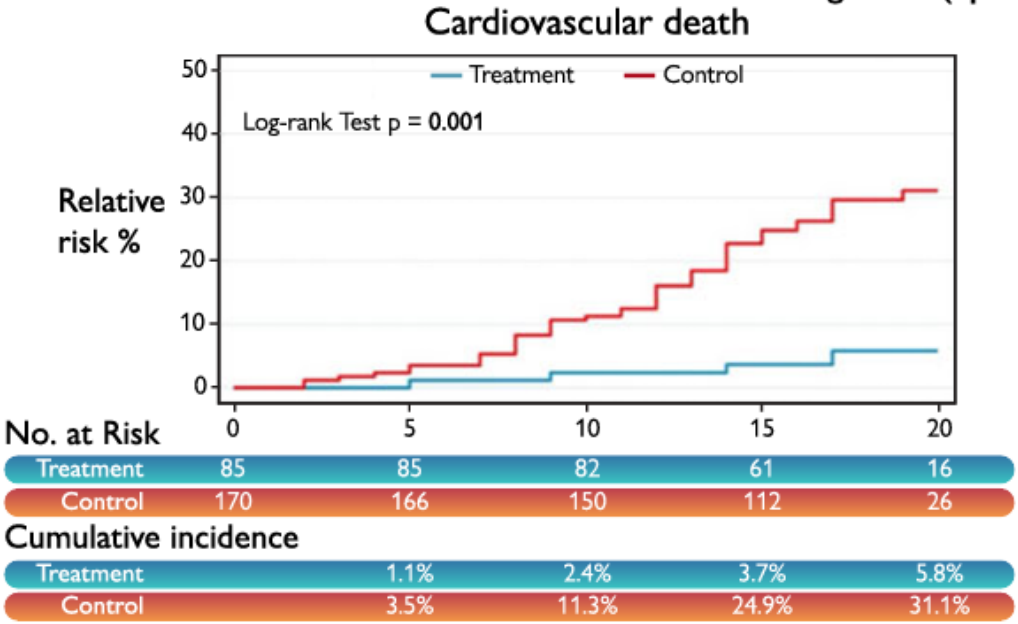
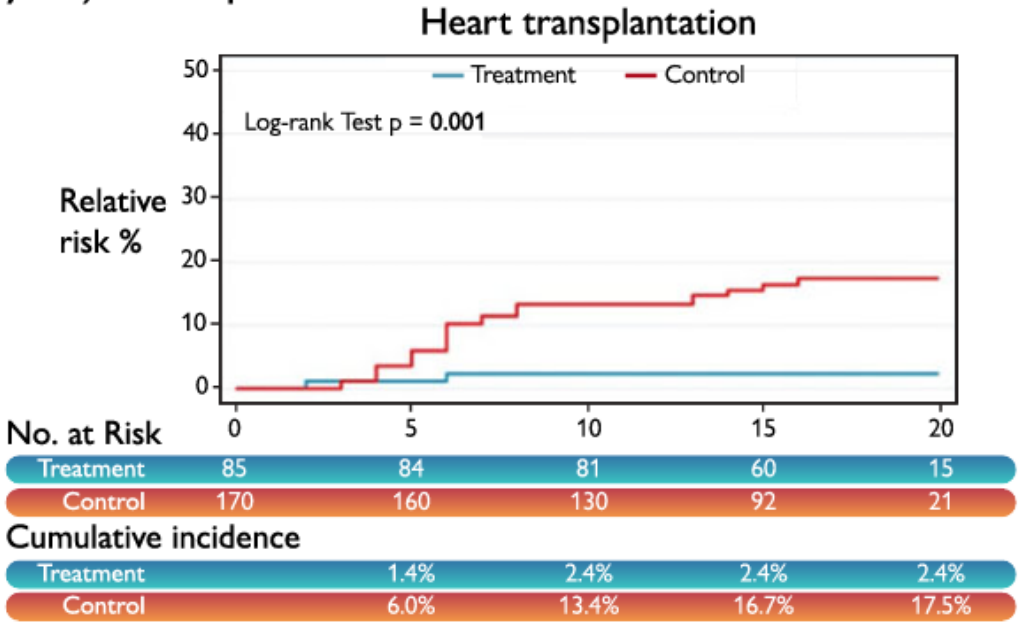
Frustaci et al., EHJ 2009
Chimenti et al. EHJ 2022

Immuunsuppressie bij inflammatoire CMP

Long-term (up to 20 years) follow-up





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Chimenti et al. EHJ 2022

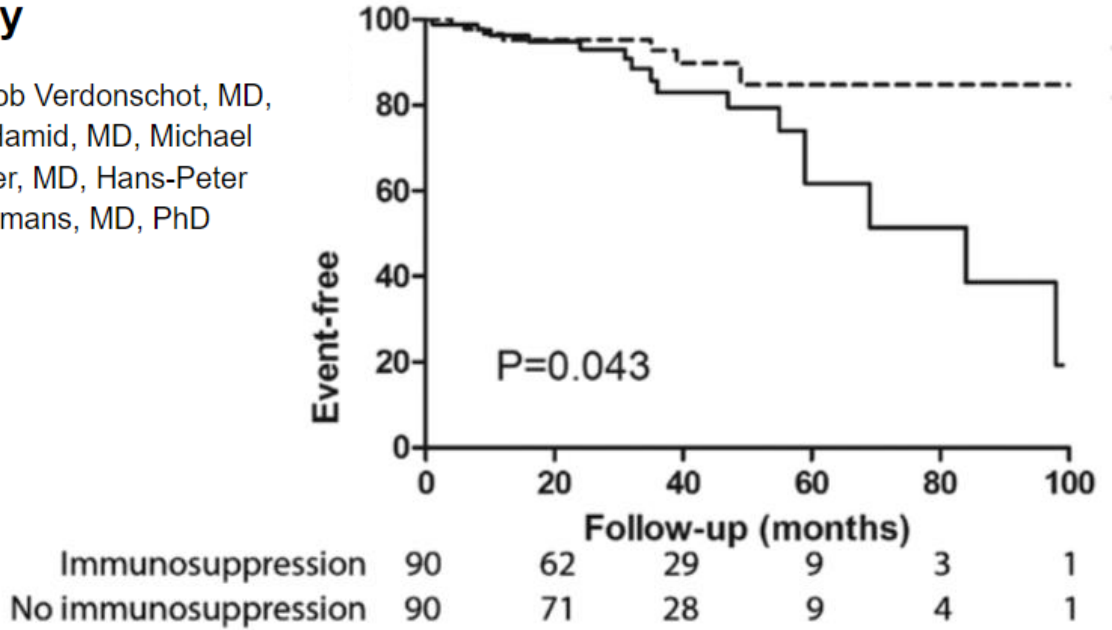
Circulation: Heart Failure
Volume 11, Issue 2, February 2018
https://doi.org/10.1161/CIRCHEARTFAILURE.117.004228

American Heart Association.

ORIGINAL ARTICLE

Immunosuppressive Therapy Improves Both Short- and Long-Term Prognosis in Patients With Virus-Negative Nonfulminant Inflammatory Cardiomyopathy

Jort Merken, MD*, Mark Hazebroek, MD*, Pieter Van Paassen, MD, Job Verdonschot, MD, Vanessa Van Empel, MD, Christian Knackstedt, MD, Myrurgia Abdul Hamid, MD, Michael Seiler, MD, Julian Kolb, MD, Philipp Hoermann, MD, Christian Ensinger, MD, Hans-Peter Brunner-La Rocca, MD, PhD, Gerhard Poelzl, MD, and Stephane Heymans, MD, PhD



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Merken et al. Circ Heart Failure 2018

Follow up traject DCM

- Patiënt-specifiek: grotendeels bepaald door ernst van aandoening, co-morbiditeiten, leeftijd en klachten

- Algemeen: Na optitratie fase controle na 6 maanden. Indien stabiel, controle na 12 maanden en indien dan stabiel, controles elke 3 jaar

Follow up traject DCM

1. DCM eci (LGE-, EMB-, genetica-):
   - CVRM via huisarts jaarlijks (m.n. nierfunctie /elektrolyten)
   - Elke 3 jaar poli + TTE bij HF cardioloog, bij (klinische) achteruitgang MRI. Holter op indicatie

2. DCM met familiale VG/genetische mutatie en/of ICD
   - Jaarlijks follow-up met poli bij HF cardioloog met DCM expertise.
   - Elke 2 jaar TTE, bij (klinische) achteruitgang MRI. Holter o.g.v. kliniek en/of aritmogene mutaties (LMNA, PLN, FLNC, DSP, RBM20)

3. DCM met EMB+
   - Policontrole in MUMC+, frequentie afhankelijk van etiologie en behandelstrategie

Zuyderland – MUMC Hartfalen team

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Maastricht UMC+

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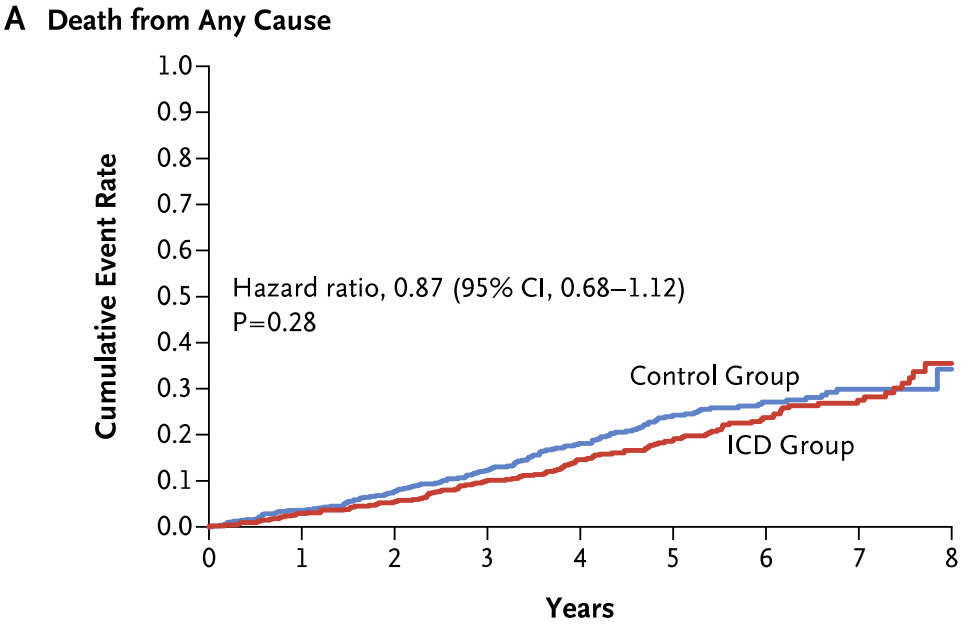
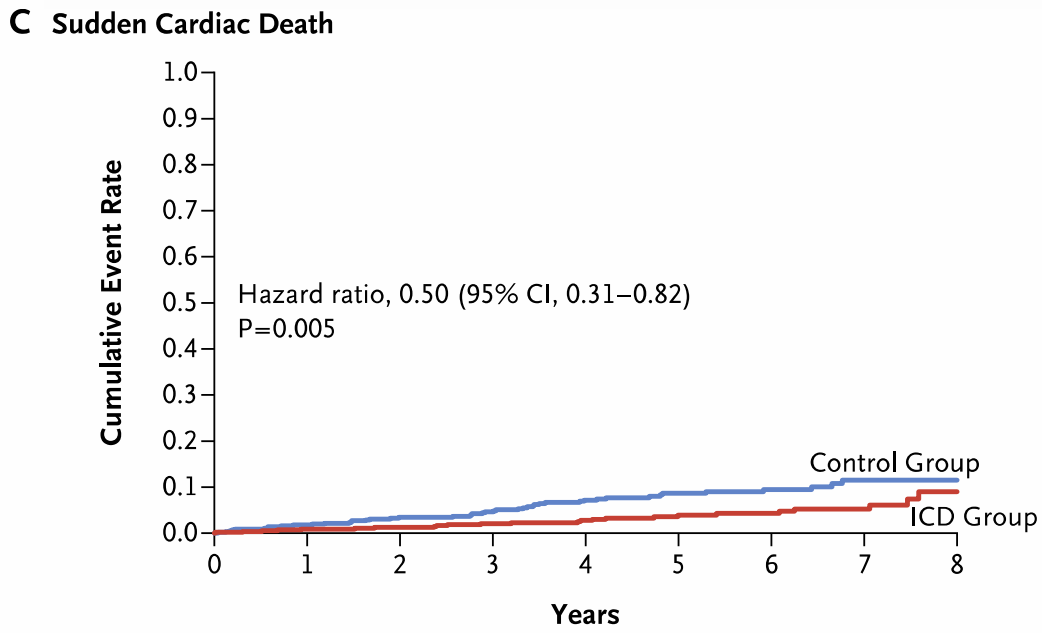
Back up slides

DANISH trial – Dilated Cardiomyopathy





- No reduction in all-cause mortality
- Low event rates for SCD
- Optimale HF medication & CRT (58%)

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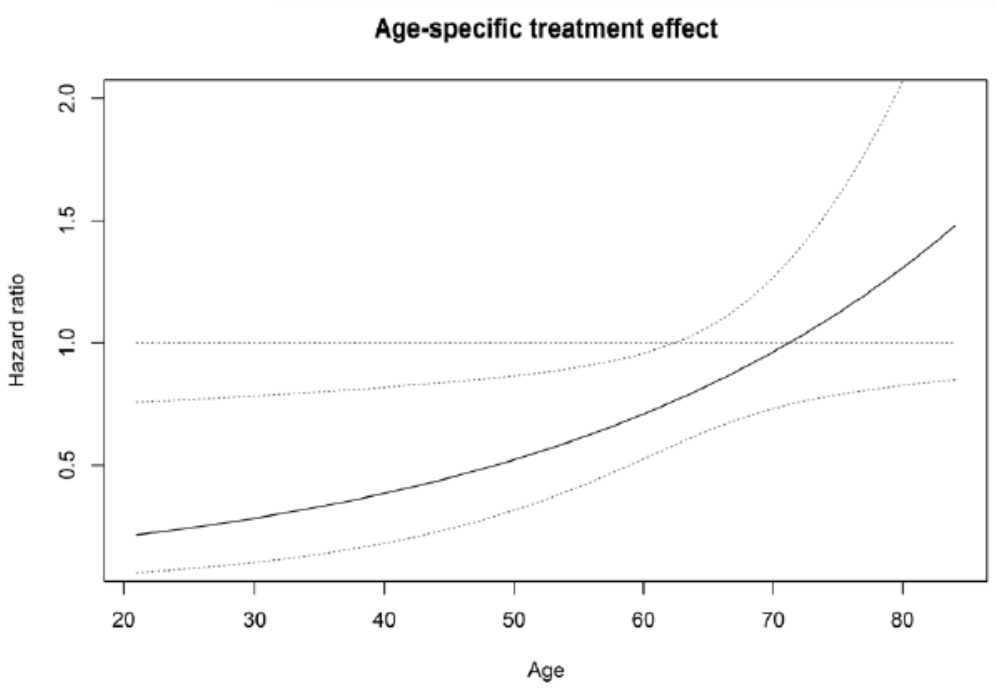
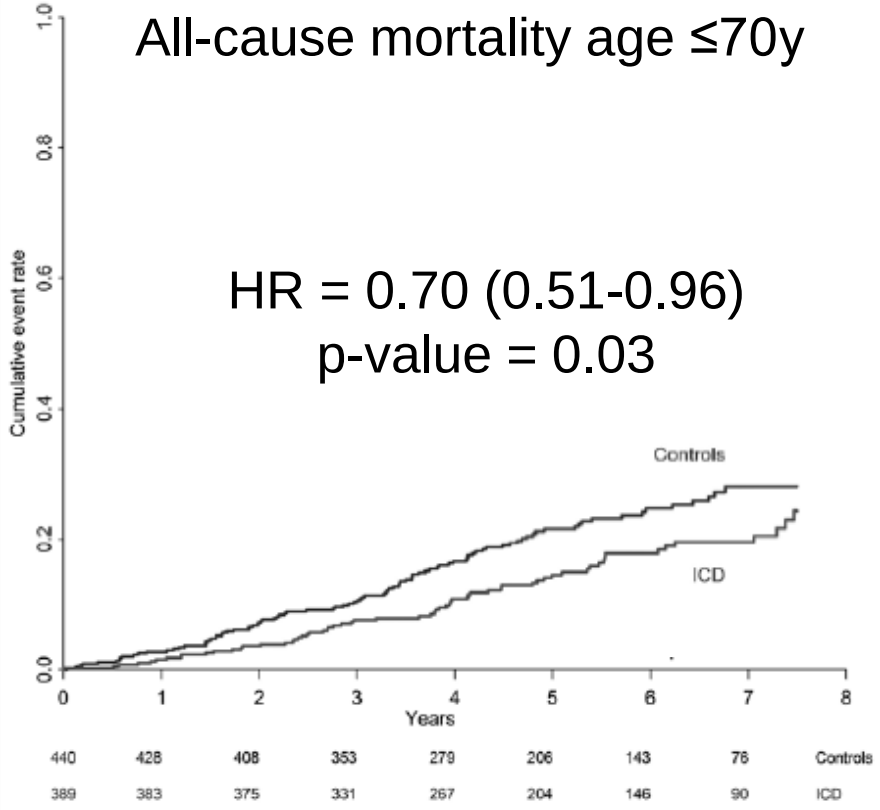
Køber et al.,NEJM 2016

Age-specific outcomes DANISH



All-cause mortality age ≤70y

HR = 0.70 (0.51-0.96)
p-value = 0.03



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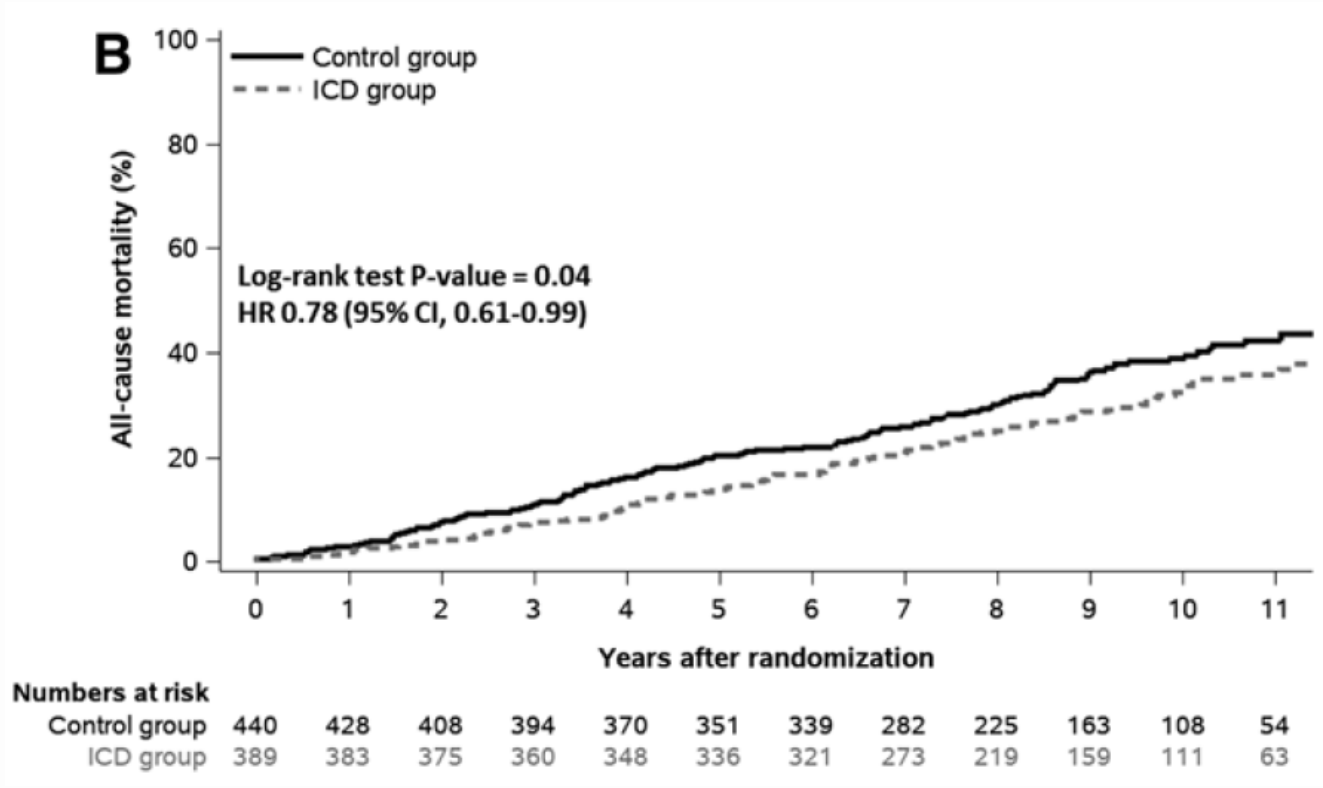
Elming et al.,Circulation 2017

DANISH long-term: all-cause mortality, <70y



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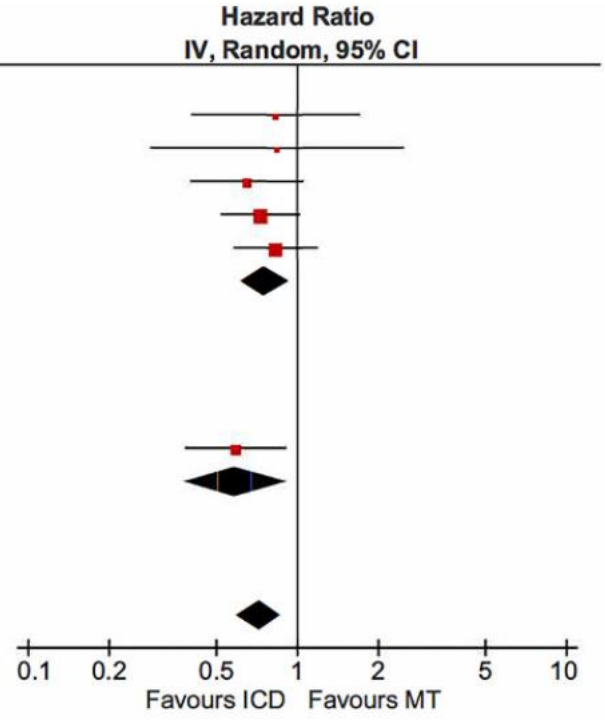
Yafasova A. et al Circ 2022

All-cause mortality reduction of 24% with ICD

Study or Subgroup	Weight	Hazard Ratio IV, Random, 95% CI	Year
3.1.1 RCT ICD-only			
CAT	6.6%	0.83 [0.40, 1.71]	2002
AMIOVIRT	2.9%	0.84 [0.28, 2.49]	2003
DEFINITE	14.7%	0.65 [0.40, 1.06]	2004
SCD-HeFT (placebo)	30.2%	0.73 [0.52, 1.02]	2005
DANISH-ICD	27.0%	0.83 [0.58, 1.19]	2016
Subtotal (95% CI)	**81.5%**	**0.76 [0.62, 0.93]**	
Heterogeneity: Tau² = 0.00; Chi² = 0.77, df = 4 (*P* = 0.94); I² = 0%			
Test for overall effect: Z = 2.64 (*P* = 0.008)			
3.1.3 Obs ICD-only			
EU-CERT-ICD	18.5%	0.59 [0.38, 0.91]	2020
Subtotal (95% CI)	**18.5%**	**0.59 [0.38, 0.91]**	
Heterogeneity: Not applicable			
Test for overall effect: Z = 2.39 (*P* = 0.02)			
Total (95% CI)	**100.0%**	**0.72 [0.60, 0.87]**	
Heterogeneity: Tau² = 0.00; Chi² = 1.82, df = 5 (*P* = 0.87); I² = 0%			
Test for overall effect: Z = 3.41 (*P* = 0.0007)			
Test for subgroup differences: Chi² = 1.04, df = 1 (*P* = 0.31), I² = 4.0%			



zuyderland

Theuns et al.,NHJ 2023