

# DGK CARDIO UPDATE 2024

16. und 17. Februar

23. und 24. Februar

## Ventrikuläre Rhythmusstörungen

Lars Eckardt, Münster

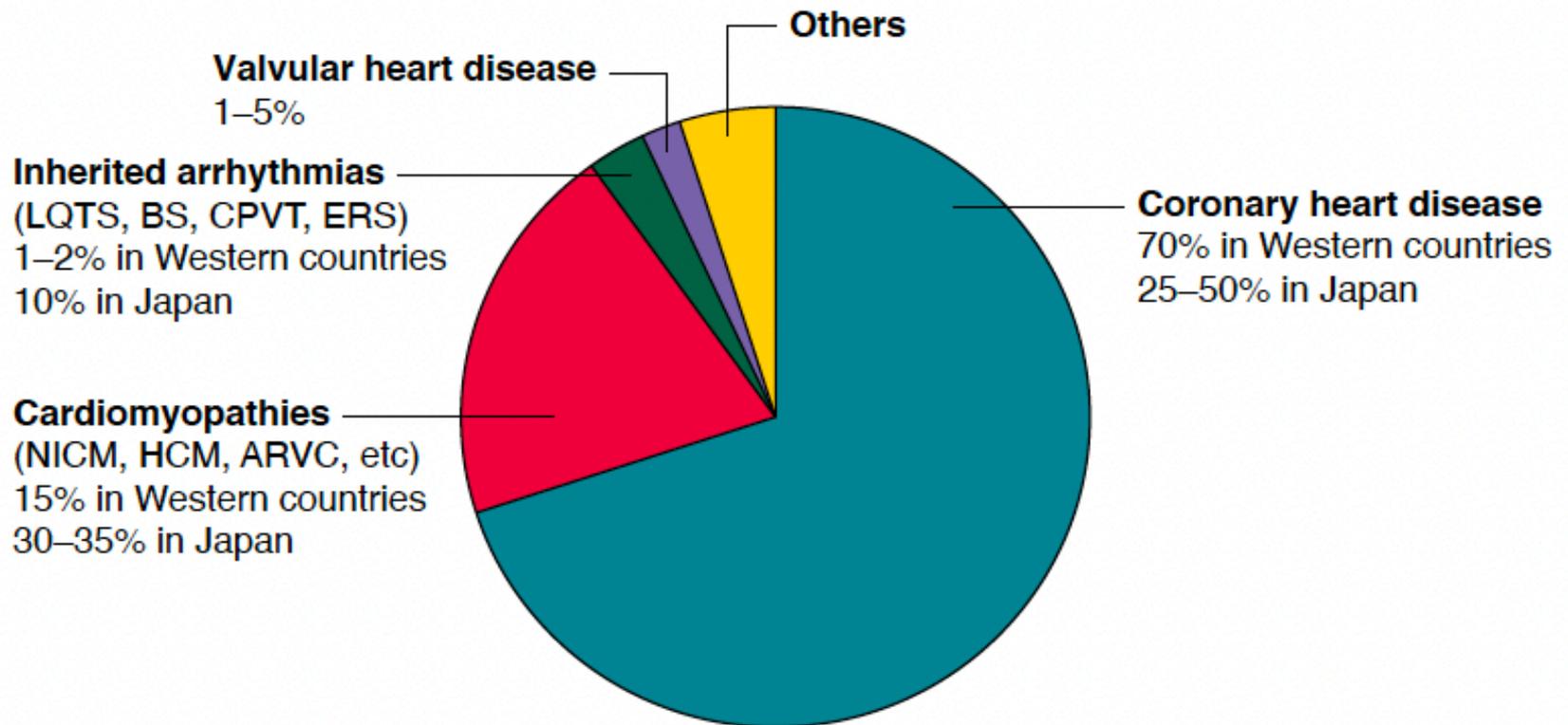
# Interessenkonflikte

<b>Research Grants</b>	Medtronic, Abbott, Biotronik, Boston Scientific, Sorin Deutsche Herzstiftung, DFG
<b>Consultant / Advisory Board</b>	Abbott, Johnson & Johnson, Biotronik; Medtronic; Novartis
<b>Ownership Interests</b>	None
<b>Speaker's Bureau Honoraria</b>	Medtronic, Abbott, Boston Scientific, Biotronik, Sanofi Aventis, Pfizer, Bristol Myers Squibb, MS, Boehringer Ingelheim, Bayer Healthcare, Daiichy Sankyo

# Neues in der Risikostratifikation Des plötzlichen Herztods

# Ursachen des plötzlichen Herztods

*Tfelt-Hansen et al., Europace 2023;25:1-9*

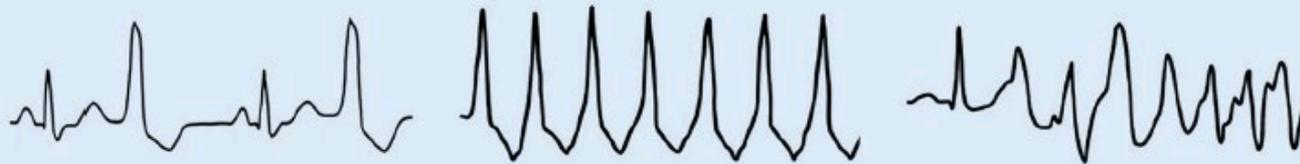


# ESC-Leitlinie: VT und SCD: Was ist neu?

*Zeppenfeld et al., Eur Heart J 2022;43:3997-4126*

*Eckardt et al., Die Kardiologie 2023;17:27–38*

Wesentliche Neuerungen der 2022 ESC-Leitlinien zum  
Management ventrikulärer Arrhythmien



Multidisziplinärer Ansatz

Kardiologie – Bildgebung – Elektrophysiologie – Notfallmedizin –  
Herzchirurgie – Genetik – Pathologie

Diagnostik

Risiko-  
stratifikation

Akuttherapie

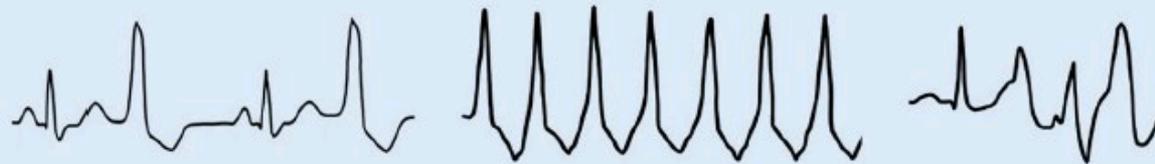
Langzeit-  
therapie

# ESC-Leitlinie: VT und SCD: Was ist neu?

*Zeppenfeld et al., Eur Heart J 2022;43:3997-4126*

*Eckardt et al., Die Kardiologie 2023;1*

## Wesentliche Neuerungen der 2022 ESC-Leitlinien zum Management ventrikulärer Arrhythmien



### Multidisziplinärer Ansatz

Kardiologie – Bildgebung – Elektrophysiologie – Notfallmedizin –  
Herzchirurgie – Genetik – Pathologie



- Individualisierte Risikostratifikation (u.a. DCM)
- Stärkere Einbeziehung genetischer Diagnostik
- Zunehmende Bedeutung der kardialen MRT
- Implementierung von Risikokalkulatoren und -markern (z.B. programmierte Stimulation)
- Pat. ohne KHK: reduzierte Aussagekraft der LVEF
- Screening von Familienangehörigen

# Personalisierte ICD-Nutzen-Berechnung

## MADIT Benefit Score (<https://is.gd/madit>)

Arthur J. Moss, Who Pioneered  
Heart Treatments, Dies at 86

Younis et al., *Eur Heart J* 2021; 42:1676-1684

Feb. 28, 2018

- Risikoabschätzung des 3 Jahres Nutzens einer primär-prophylaktischen ICD-Versorgung anhand aller Pat. der MADIT Studien (1997-2011)

Charakteristika	4531
Medianes Alter	64 J
Frauen	24%
CRT-D	40%
Mediane LVEF	25±6%
ICM	66%
NSVT	7%

# Personalisierte ICD-Nutzen-Berechnung

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Charakteri

Medianes A

Frauen

CRT-D

Mediane LV

ICM

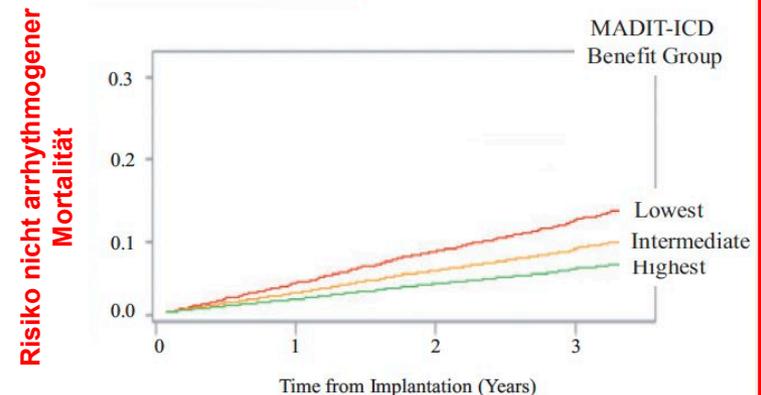
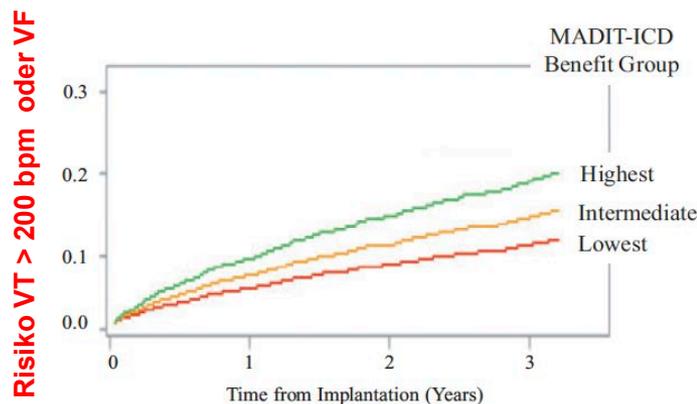
NSVT

MADIT-ICD Benefit Group and corresponding personalized ICD-Benefit Score

MADIT-ICD Benefit Group	Lowest	Intermediate	High
VT/VF Score	Low (<7)	Low (<7)   High (≥7)	High (≥7)
Non-arrhythmic Mortality Score	High (≥3)	Low (<3)   High (≥3)	Low (<3)
ICD-Benefit Score	0	13 25 38 50 63 75 88	100

VT/VF Score	
Variable	Points
LVEF ≤25%	+1
Atrial arrhythmia	
Heart Rate > 75 bpm	
SBP <140 mmHg	+2
Myocardial Infarction	
Age < 75 yrs	
Male	
Prior NSVT	

Non-arrhythmic Mortality Score	
Variable	Points
CRT-D	-1
NYHA ≥II	+1
Diabetes	
BMI <23 kg/m <sup>2</sup>	+2
Atrial arrhythmia	
LVEF ≤25%	
Age ≥ 75 years	

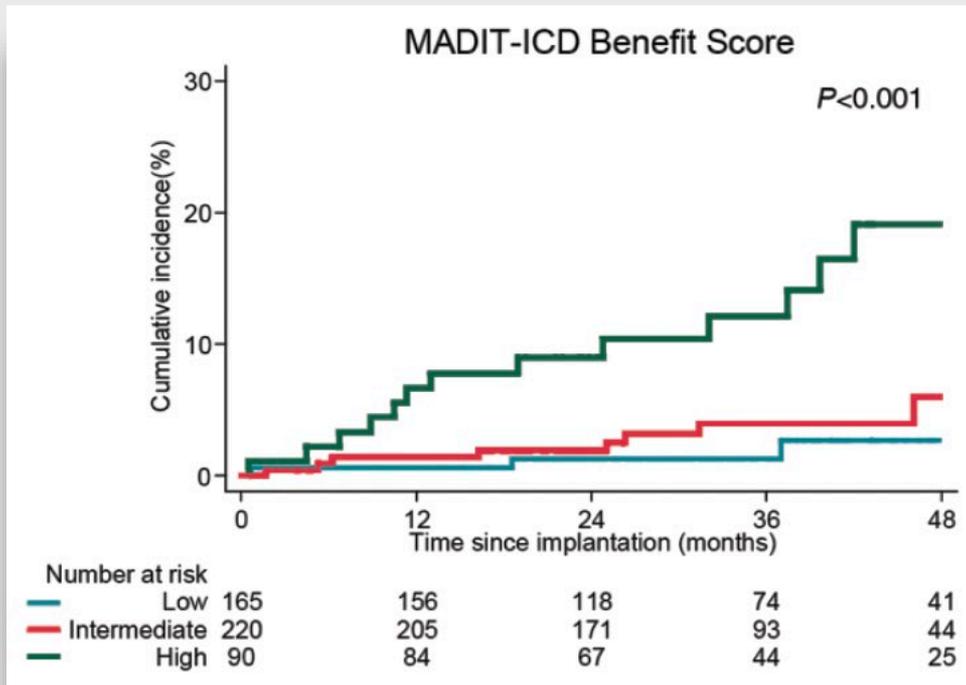


# Personalisierte ICD-Nutzen-Berechnung auch für CRT

*Dauw et al., Europace 2022; 24:1276–1283*

Retrospektive Analyse von 475 Pat. (LVEF 26%; ICM 40%; NYHA II/III;  
*Benefit Score*: 35%, 46%, 19% “lowest/intermediate/highest”

**Endpunkte:** anhaltende VT/VF und nicht rhythmogener Tod über 3 Jahre



MADIT ICD Score	VT/VF	Nicht rhythmogener Tod
Low	2%	19%
Intermediate	4%	15%
High	14%	3%

*Score hilfreich für die Entscheidung CRT-D vs. CRT-P!*

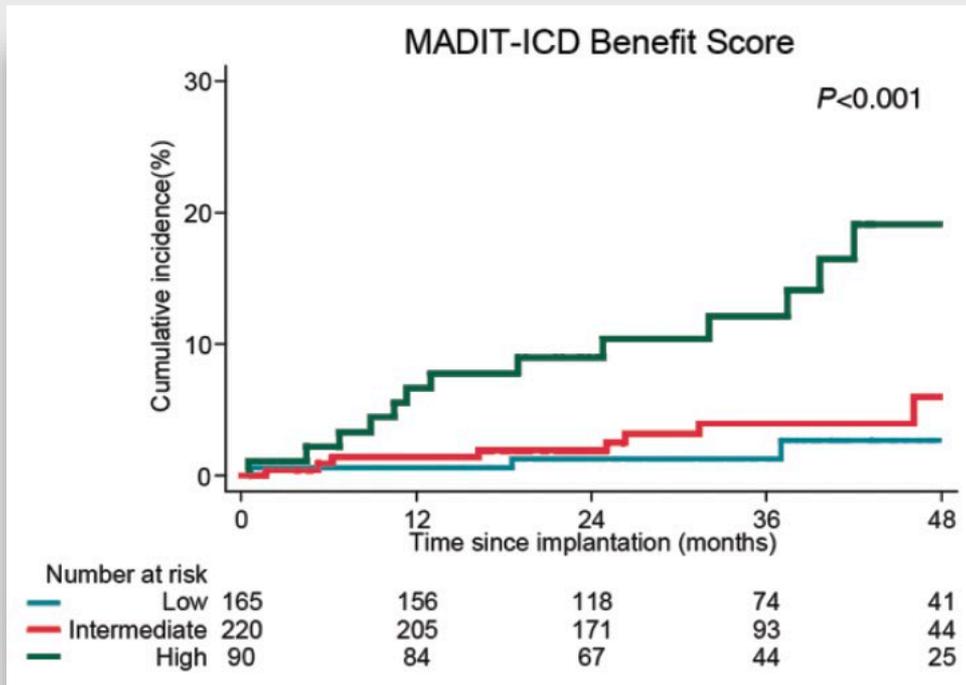
# The MADIT-ICD benefit score helps to select implantable cardioverter-defibrillator candidates in cardiac resynchronization therapy

# en-Berechnung

Dauw et al., *Europace* 2022; 24:1276–1283

Retrospektive Analyse von 475 Pat. (LVEF 26%; ICM 40%; NYHA II/III;  
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*Score hilfreich für die Entscheidung CRT-D vs. CRT-P!*

# ESC-Leitlinie: Aufwertung der kardialen MRT

*Zeppenfeld et al., Eur Heart J 2022;43:3997-4126*

*Eckardt et al., Die Kardiologie 2023;17:27–38*



## Klasse I Empfehlungen zur kardialen MRT-Diagnostik

	Empfehlung der ESC-Leitlinie 2022
Überlebende eines Herzstillstands ohne eindeutige Ursache	I
Vermutete arrhythmogene rechtsventrikuläre Kardiomyopathie	I
Hypertrophe Kardiomyopathie	I
Sportler mit auffälliger Anamnese, körperlicher Untersuchung und/oder EKG-Auffälligkeiten	I

# ESC-L

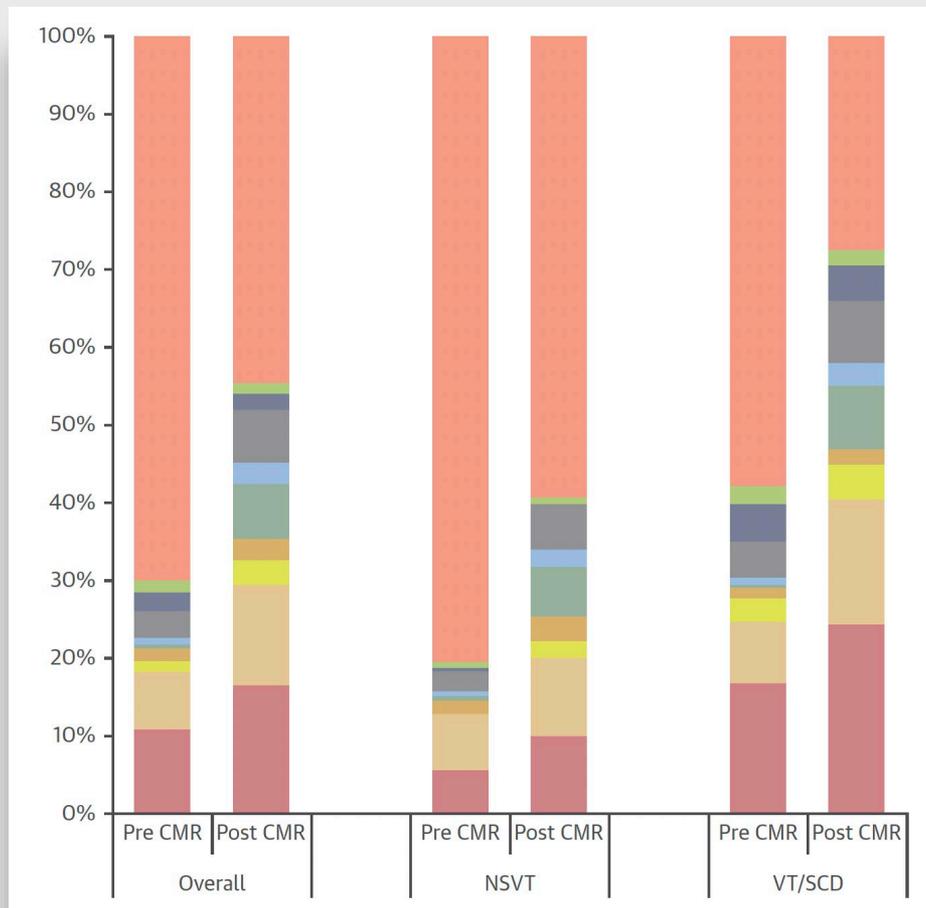
## Klasse I

Überlebend
Vermutete a Kardiomyop
Hypertrophe
Sportler mit und/oder Ek

Tab. 1 Klasse-I- und -IIa-Empfehlungen für kardiale Magnetresonanztomographie (MRT)	
	Empfehlungsgrad
<i>Allgemein</i>	
Überlebende eines HerzKreislaufstillstands ohne klare zugrunde liegende Ursache	I
Erstdokumentation einer VA (häufige VES, nsVT, monomorphe VT) und Verdacht auf eine strukturelle Herzerkrankung außer KHK	IIa
Verwandte eines am „sudden arrhythmic death syndrome“ Verstorbenen	IIb
<i>Idiopathische VES/VT und VES-induzierte Kardiomyopathie</i>	
Patienten mit nicht typischen idiopathischen VES/VT trotz unauffälliger Echokardiographie	IIa
Verdacht auf VES-induzierte Kardiomyopathie	IIa
<i>Strukturelle Herzerkrankung</i>	
Verdacht auf arrhythmogene rechtsventrikuläre Kardiomyopathie	I
Hypertrophe Kardiomyopathie	I
Dilatative Kardiomyopathie/hypokinetische nicht dilatative Kardiomyopathie	IIa
<i>Spezifische Patientengruppen</i>	
Sportler mit auffälliger Anamnese, auffälligen Befunden der körperlichen Untersuchung oder EKG-Veränderungen	I
<i>EKG</i> Elektrokardiographie, <i>KHK</i> koronare Herzkrankheit, <i>VA</i> ventrikuläre Arrhythmie, <i>VT</i> ventrikuläre Tachykardie, <i>nsVT</i> nicht anhaltende VT, <i>VES</i> ventrikuläre Extrasystolen	

# Bedeutung der kardialen MRT-Diagnostik bei Pat. mit Kammertachykardien

Ge et al., JACC CI 2023;16:1536-1549



642 Pat. (345 nsVT; 297 VT/VF)  
4 Jahre Follow-up  
Mediane LV-EF 58%  
MACE: 15% (nsVT); 35% VT/VF

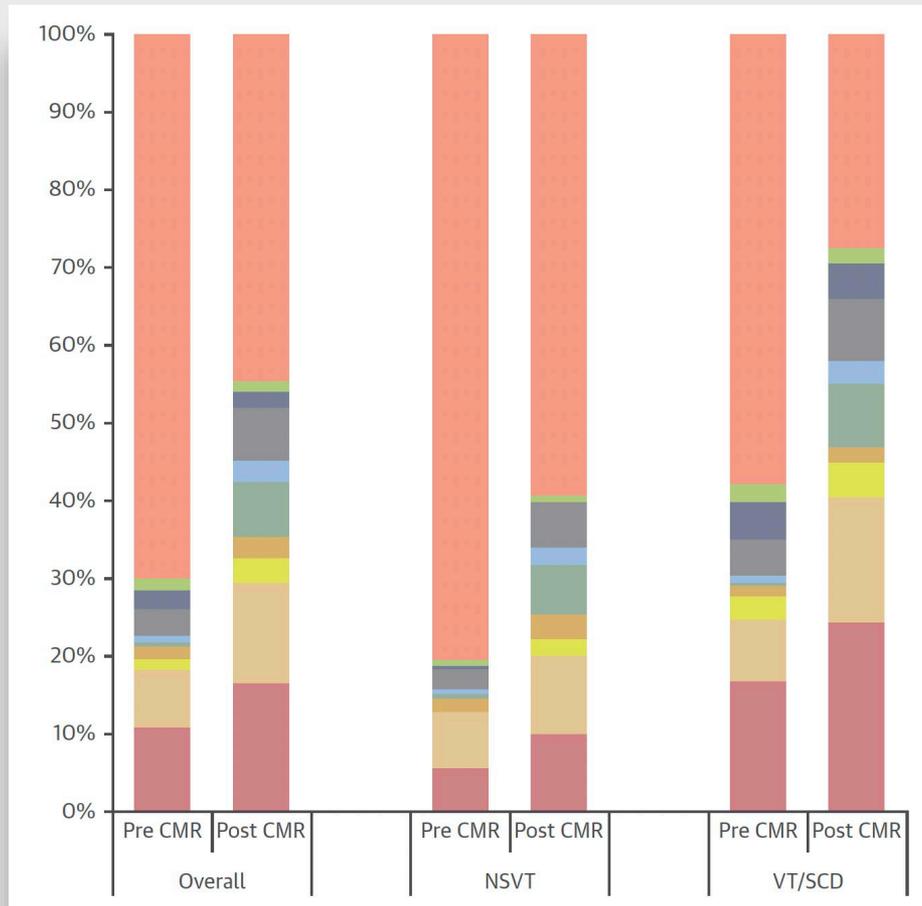
- Idiopathic
- Non-Cardiac
- Channelopathies
- Other Structural Myocardial Abnormalities
- Sarcoidosis
- Suspected Myocarditis
- HCM
- ARVC
- NICM
- ICM

# Diagnostic Impact and Prognostic Value of Cardiac Magnetic Resonance in Patients With Ventricular Arrhythmias



# T-Diagnostik ykardien

Ge et al., JACC CI 2023;16:1536-1549



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- Suspected Myocarditis
- HCM
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- NICM
- ICM



# Herzinsuffizienz – Klinik – kardiales MRT

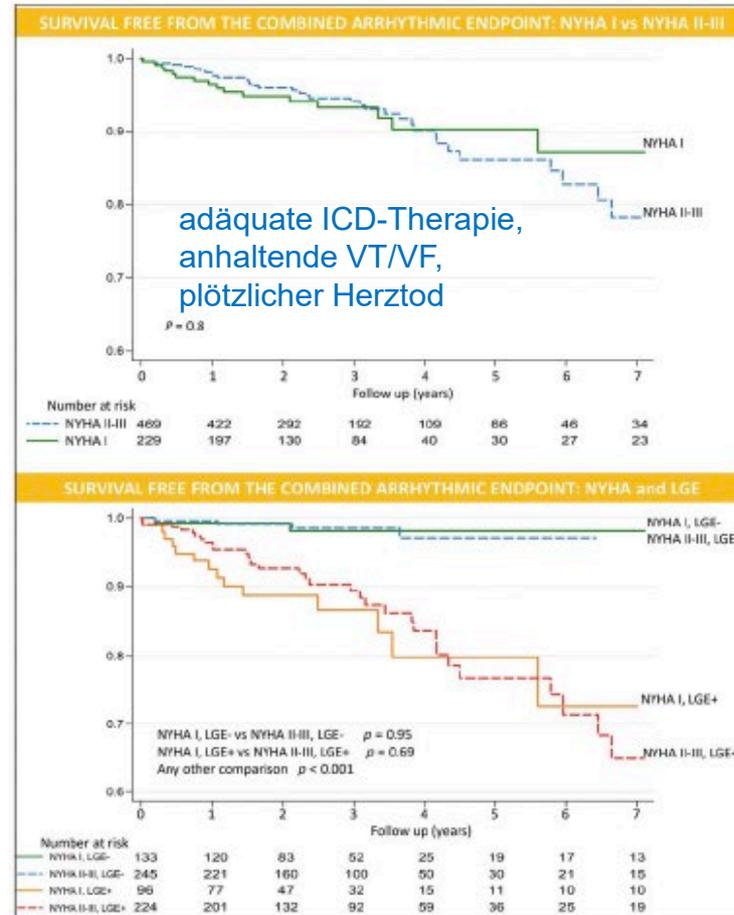
Di Marco et al., Eur J Heart Failure 2023;25:740-750

698 patients with NICM, 229 (33%) in NYHA I,  
Median follow-up 31 months.

The risk of ventricular arrhythmias and sudden death is similar between NYHA I and NYHA II-III patients ( $p=0.8$ ).

LGE is a significant discriminator of the arrhythmic risk both in NYHA I and in NYHA II-III cases.

LGE+ NYHA I patients have a considerable risk of ventricular arrhythmias and sudden death, significantly higher than LGE- NYHAII-III cases ( $p<0.001$ ).



# Late gadolinium enhancement and the risk of ventricular arrhythmias and sudden death in NYHA class I patients with non-ischaemic cardiomyopathy

# Technik – kardiales MRT

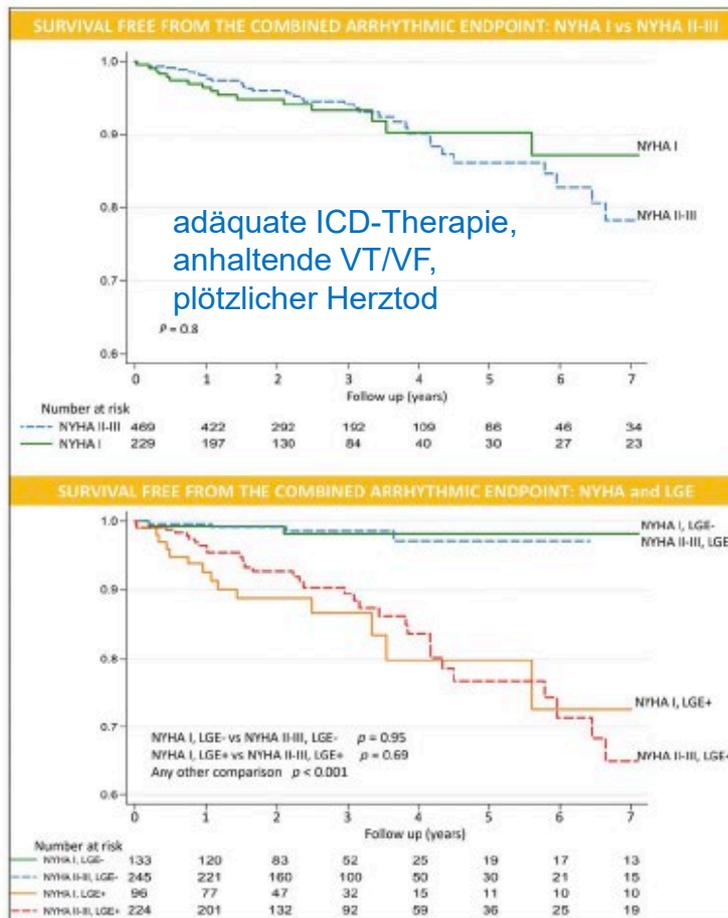
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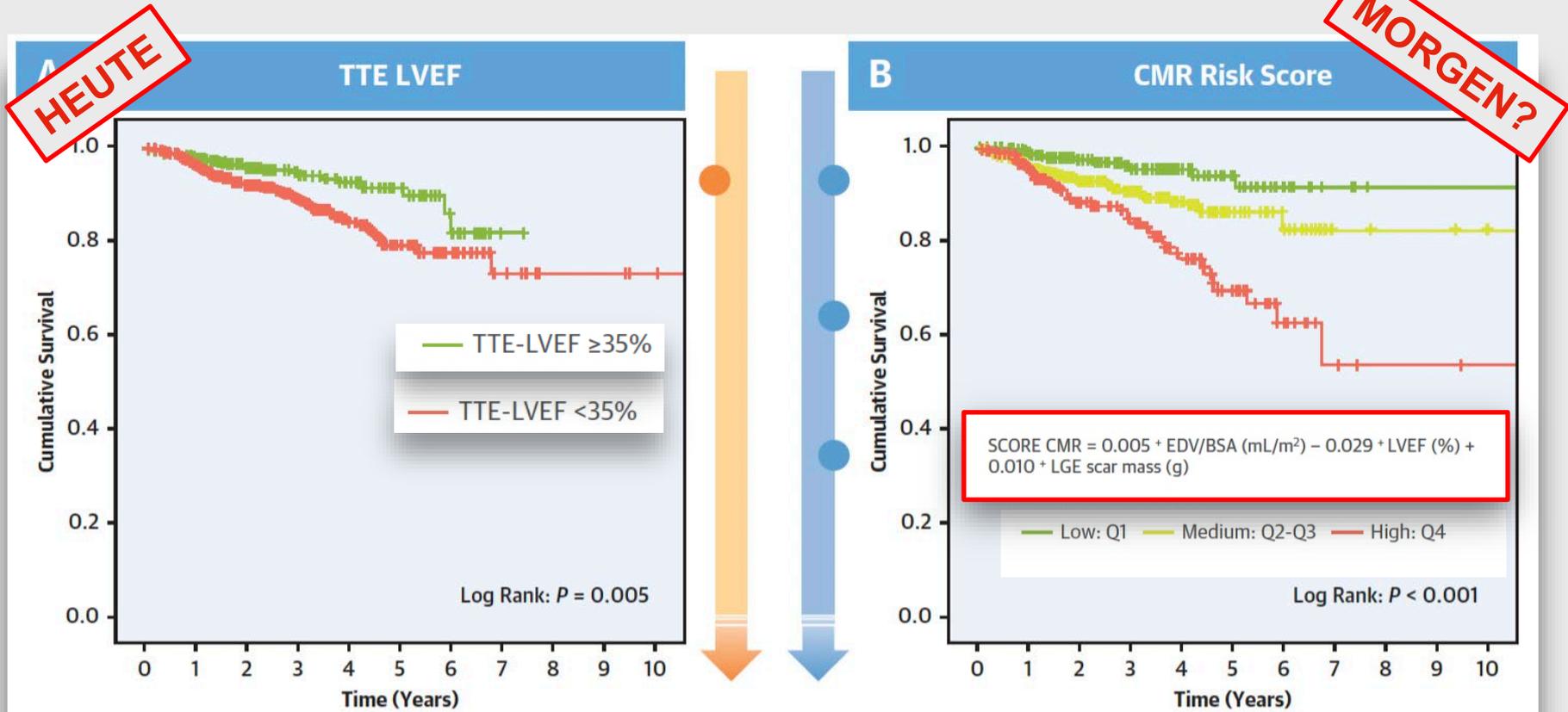


# Echo versus MRT

DERIVATE Register

Pontone G et al., JACC CI 2023;16:1387-1400

861 ICM Pat. TTE-LVED <50%; MAACE 10% (3 Jahre): 10% in ca.3 Jahren



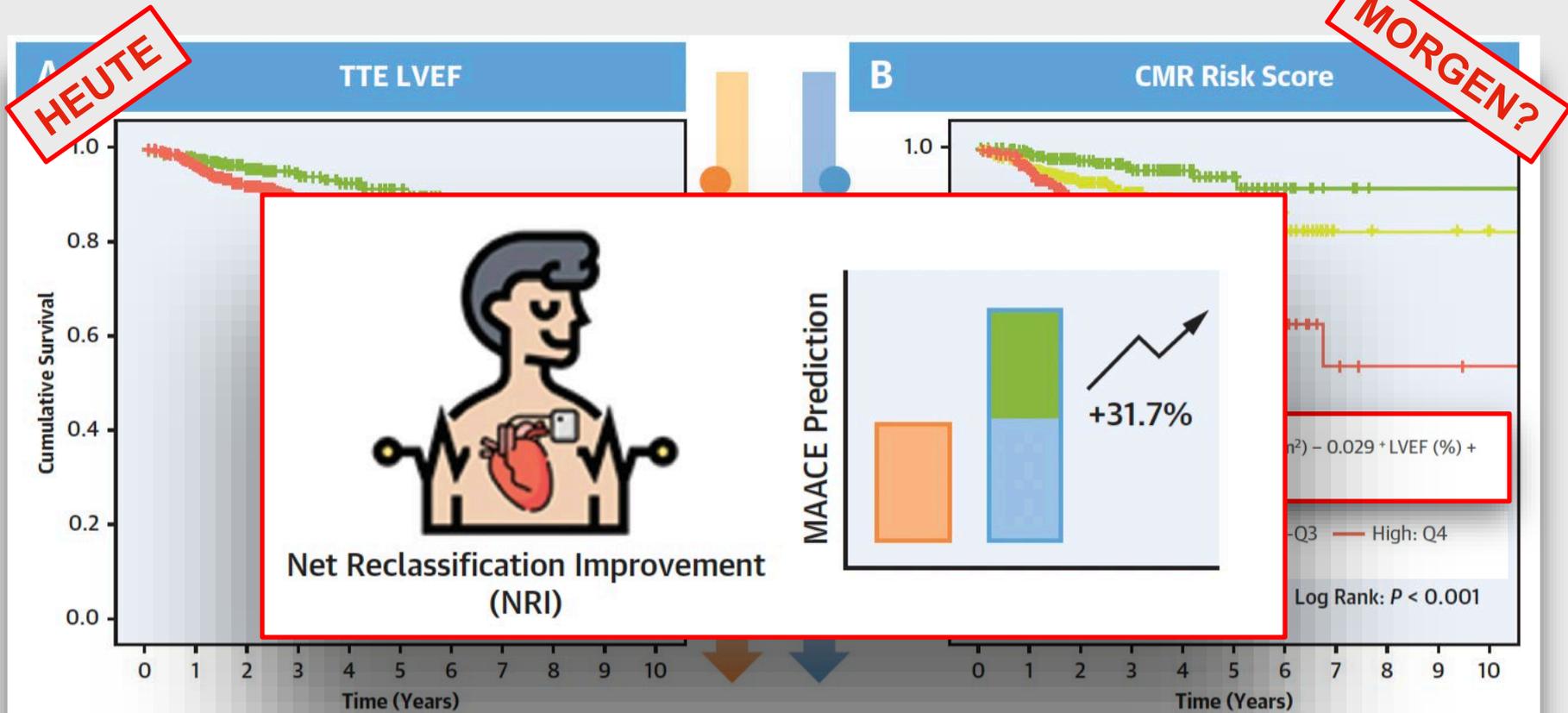
# Cardiac Magnetic Resonance for Prophylactic Implantable-Cardioverter Defibrillator Therapy in Ischemic Cardiomyopathy

## Bus MRT

**DERIVATE Register**

Pontone G et al., JACC CI 2023;16:1387-1400

861 ICM Pat. TTE-LVED <50%; MAACE 10% (3 Jahre): 10% in ca.3 Jahren

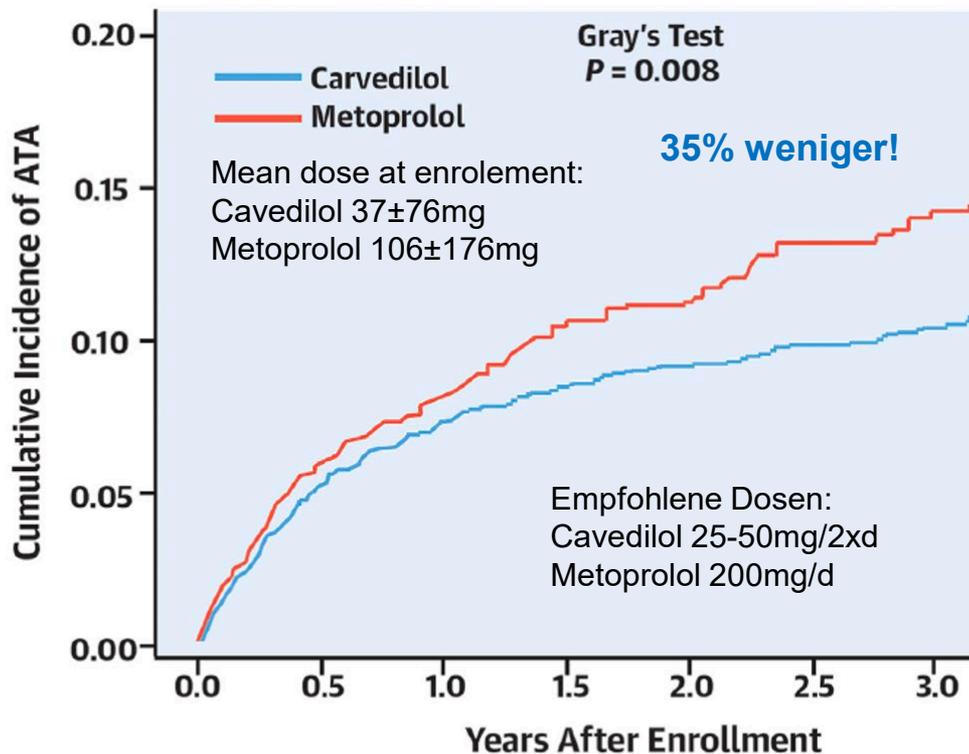


**Neues in der (med.)  
Therapie:  
Elektrischer Sturm etc.**

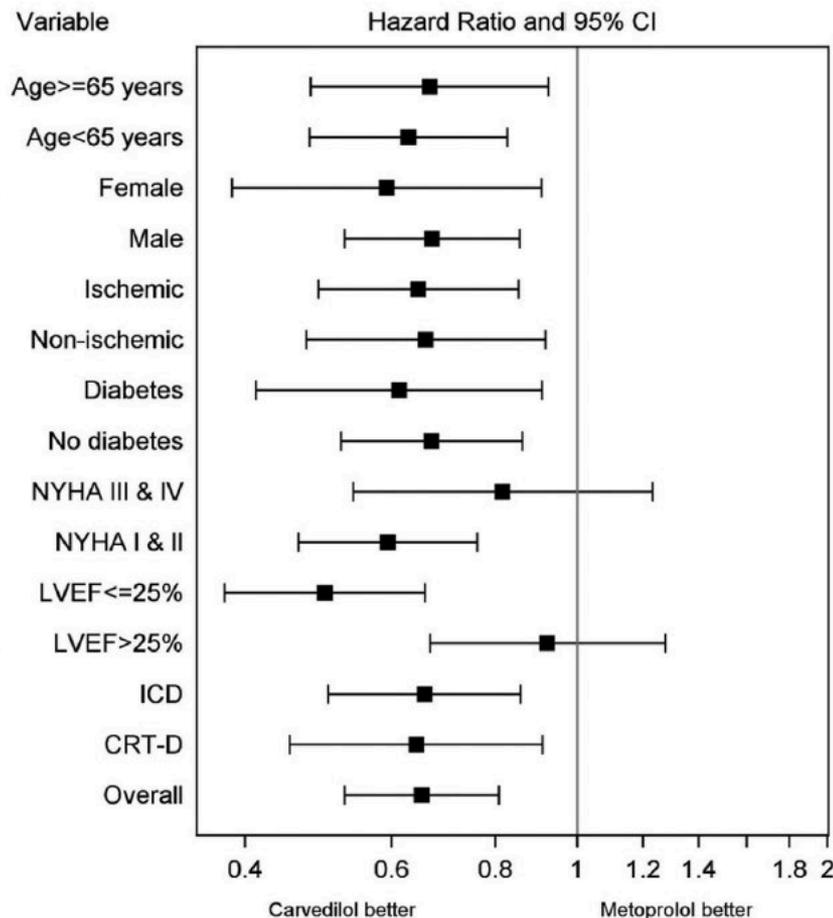
# VT-Reduktion: welcher Beta-Blocker?

Diamond et al., JACC CE 2023; 9:2122-2131

Feb. 28, 2018



Number of Patients at Risk						
2,920	2,558 (.05)	2,223 (.07)	1,796 (.08)	1,467 (.09)	1,163 (.10)	866 (.11)
1,274	1,102 (.06)	958 (.08)	770 (.11)	636 (.11)	501 (.13)	354 (.14)



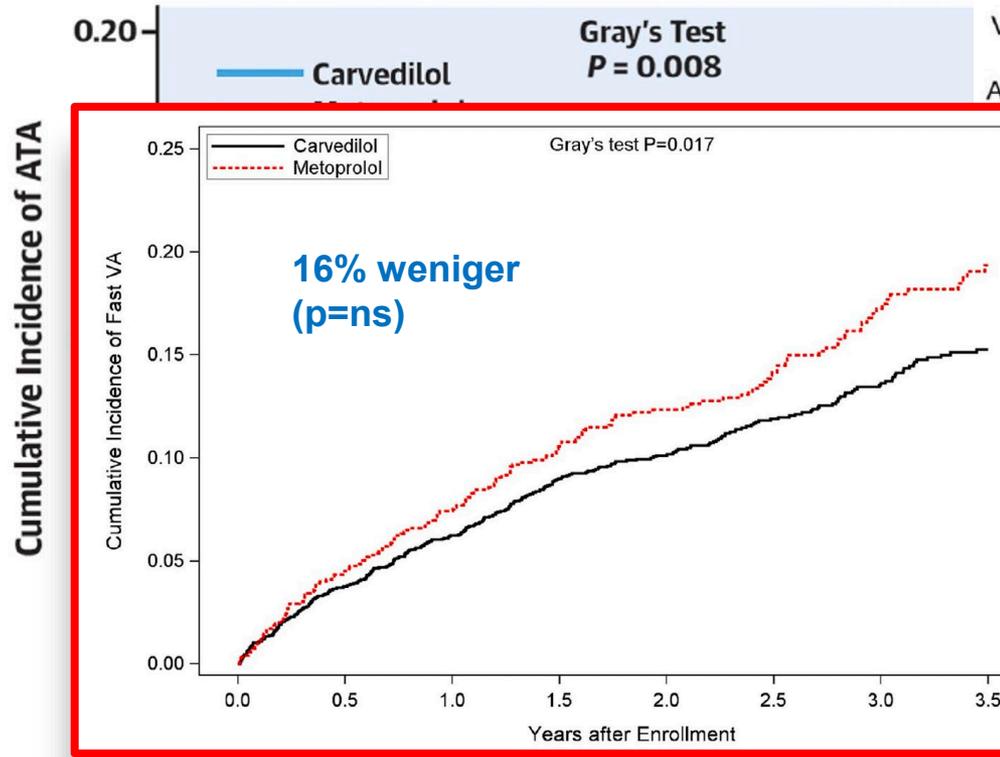
Effect of Carvedilol vs Metoprolol on Atrial and Ventricular Arrhythmias Among Implantable Cardioverter-Defibrillator Recipients

Arthur J. Moss, Who Pioneered Heart Treatments, Dies at 86

# er Beta-Blocker?

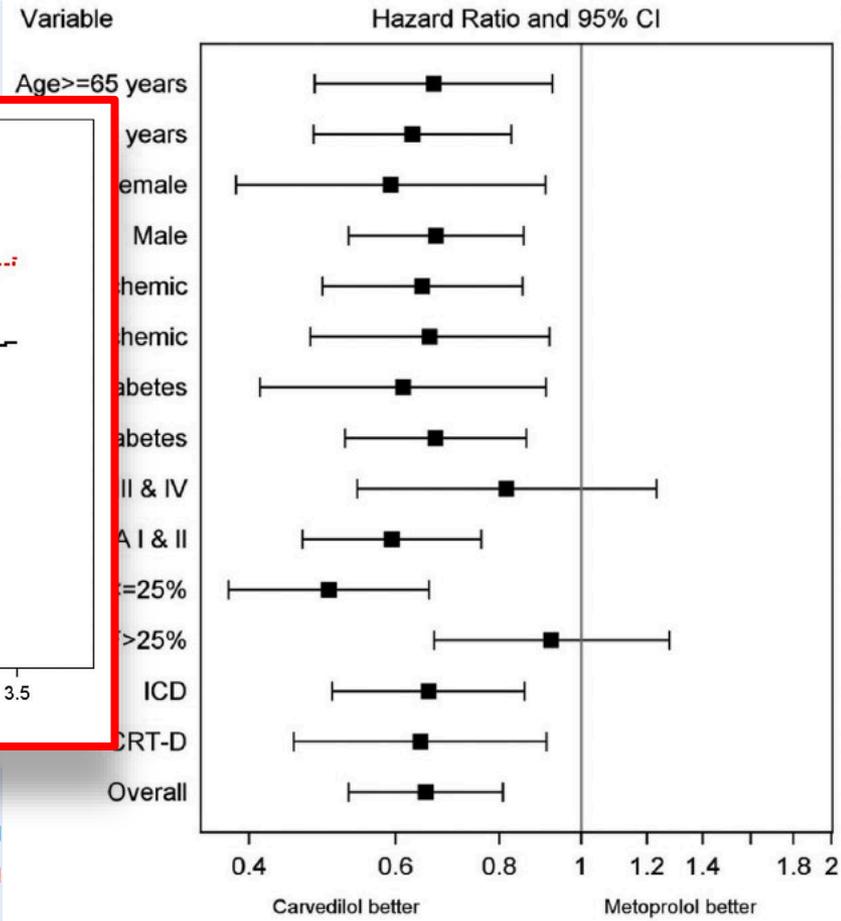
Diamond et al., JACC CE 2023; 9:2122-2131

Feb. 28, 2018



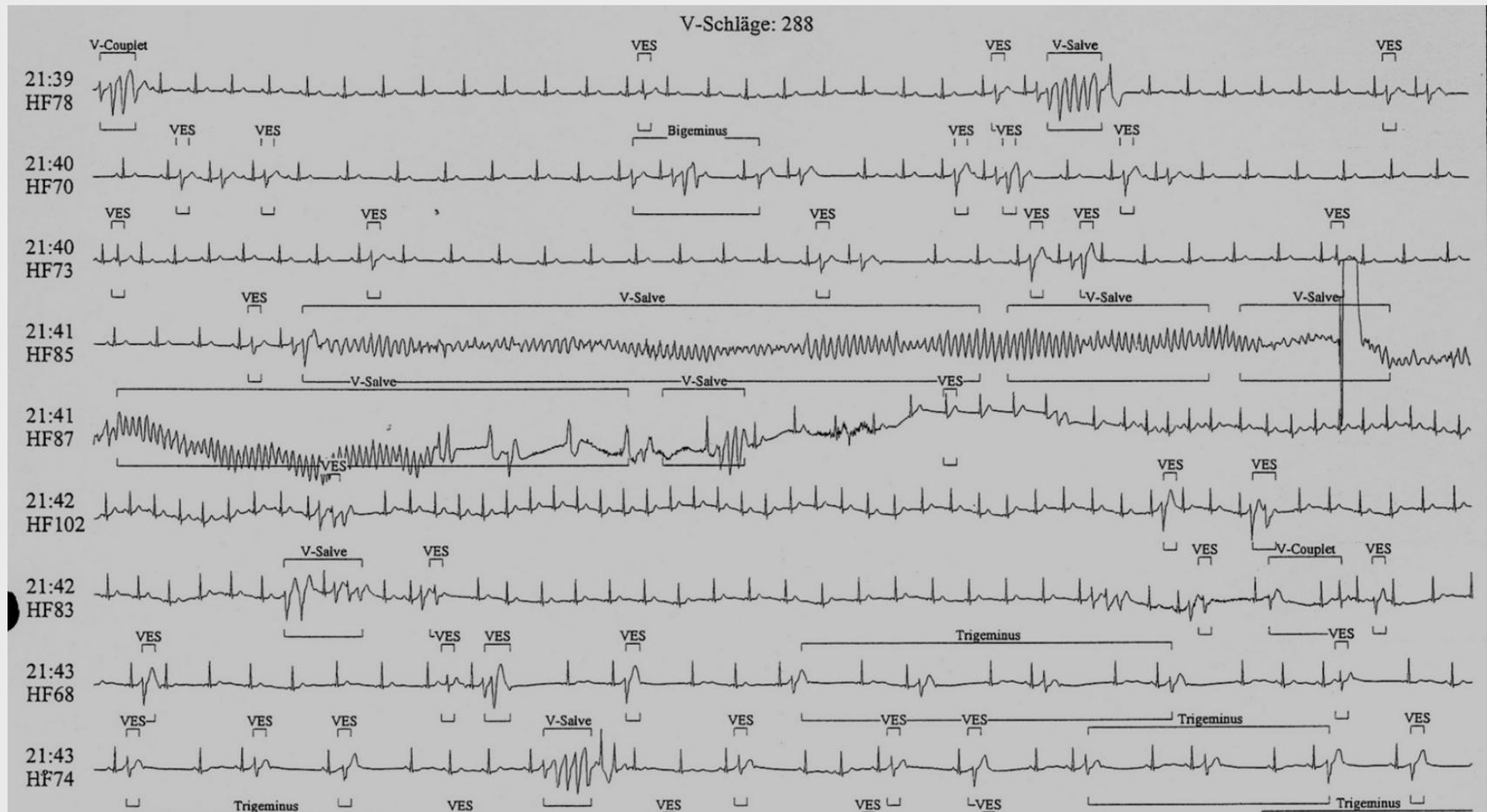
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# Elektrischer Sturm: State of the Art

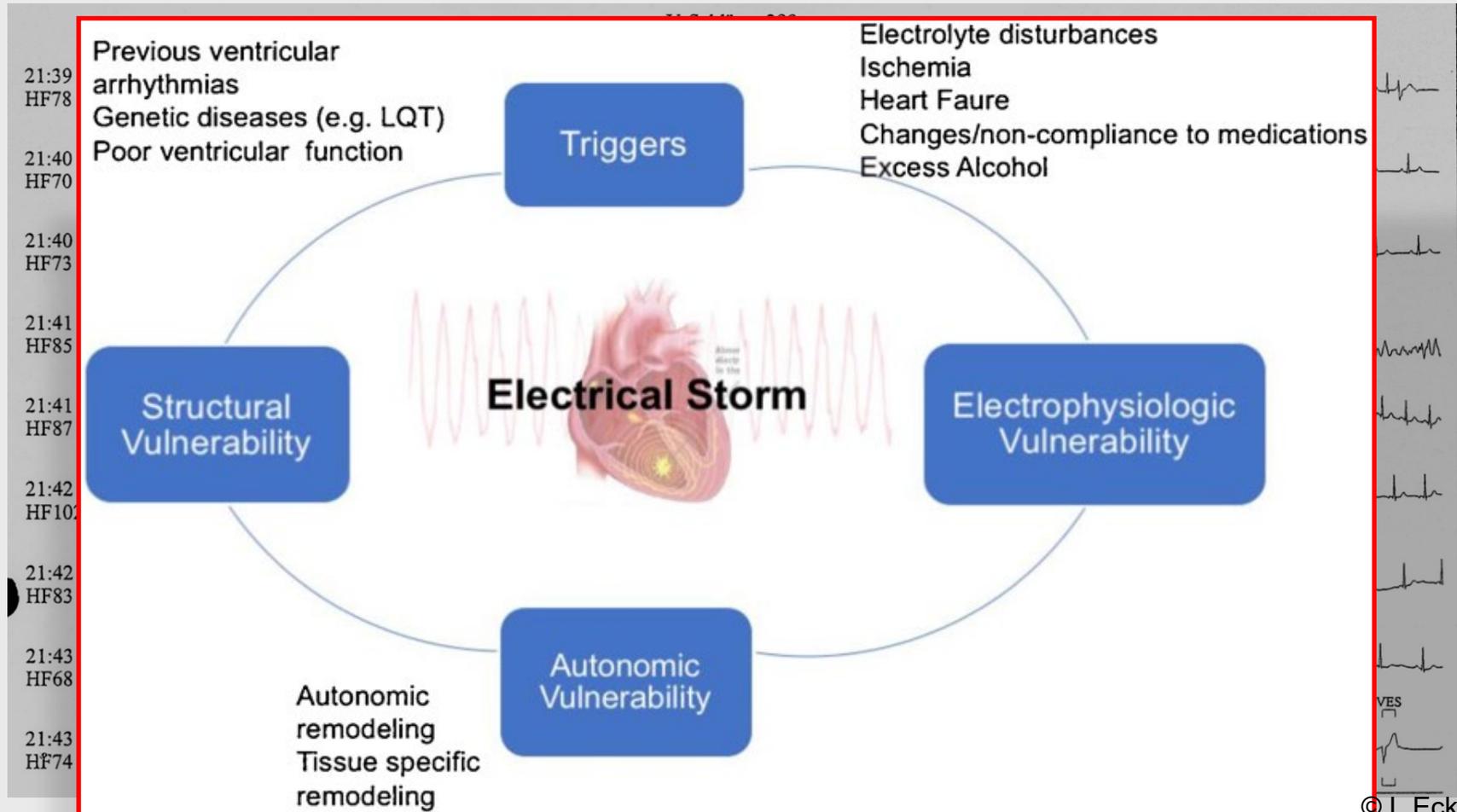
*Geraghty et al., Heart Lung Circ 2019;28:123-133*



© L.Eckardt

# Elektrischer Sturm: State of the Art

Geraghty et al., Heart Lung Circ 2019;28:123-133



# ESC-Leitlinie: VT und SCD: Was ist neu?

*Zeppenfeld et al., Eur Heart J 2022;43:3997-4126*

*Eckardt et al., Die Kardiologie 2023;17:27–38*



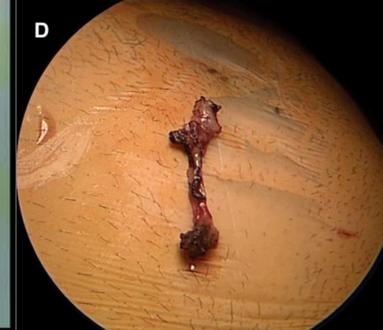
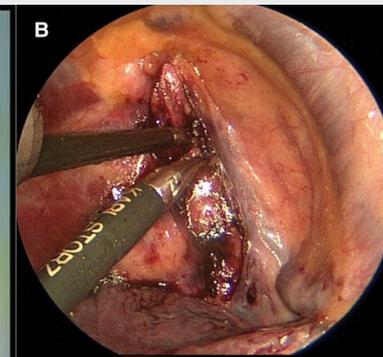
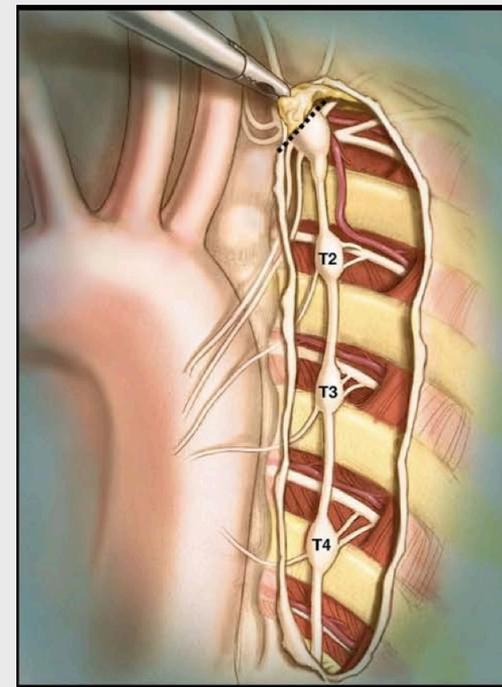
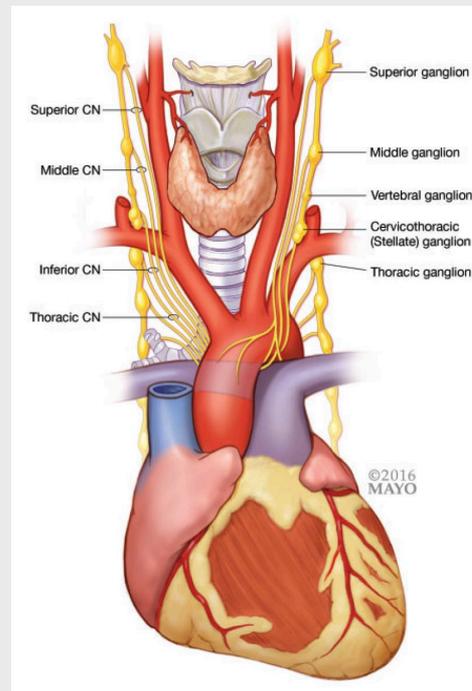
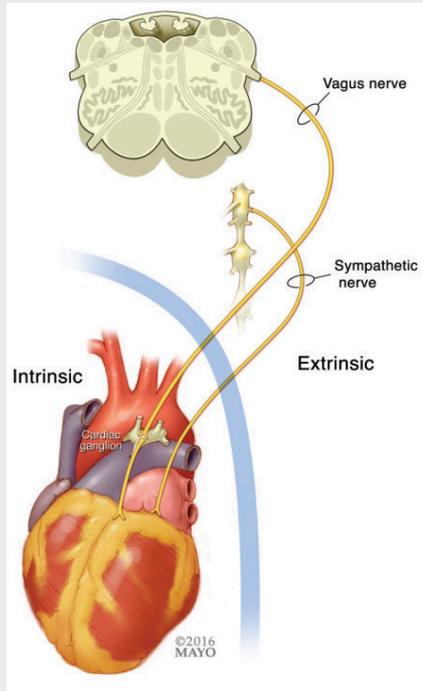
Modulation des autonomen Nervensystems kann bei Pat. mit

- Medikamenten refraktärem elektrischen Sturm oder
- Ineffektivität einer Ablation oder
- falls diese nicht möglich,

erwogen werden.

# Thoroskopische Sympathikus Denervierung

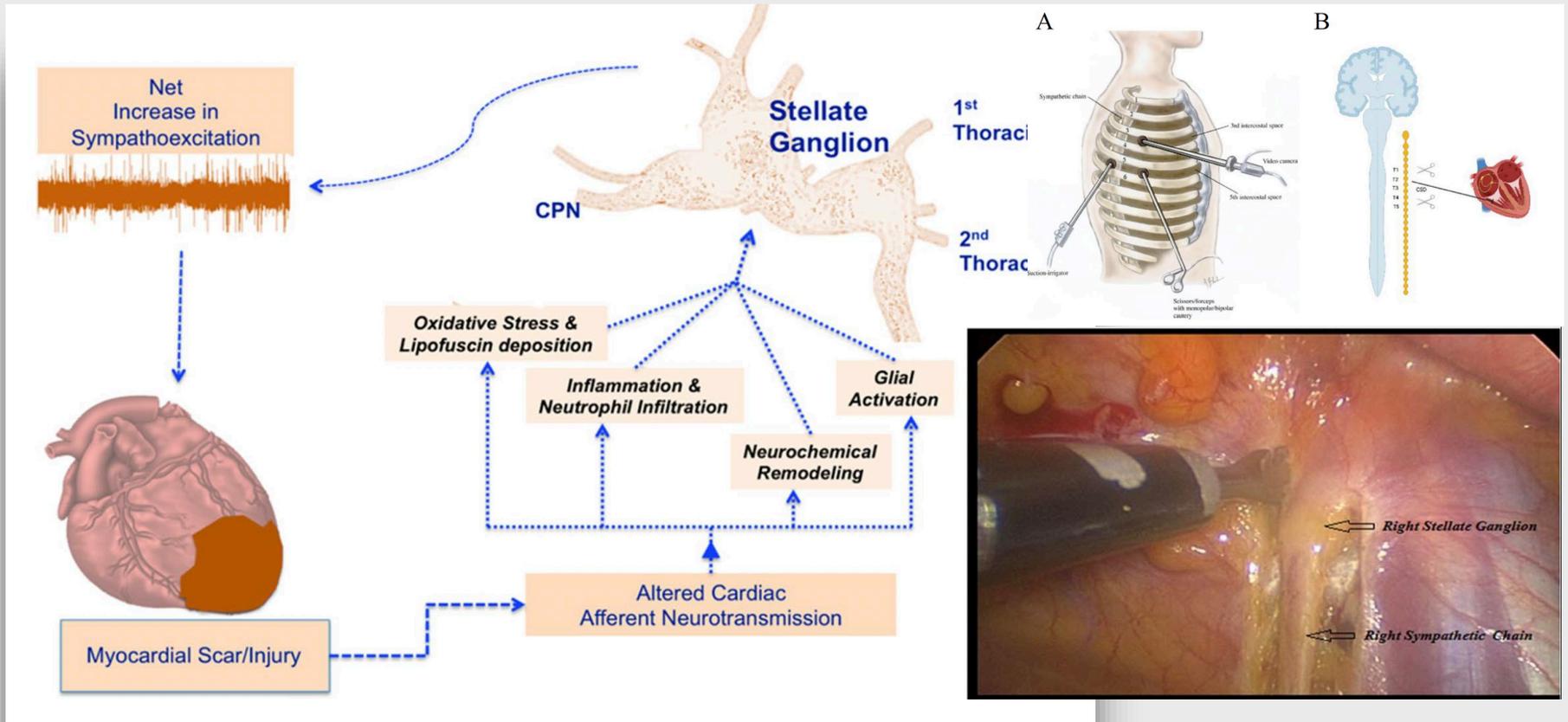
Collura et al., *Heart Rhythm* 2009; 24; 1800; König et al., *Europace* 2022;24:1800-1808



Witt et al., *Europace* 2017;19:1075-1083

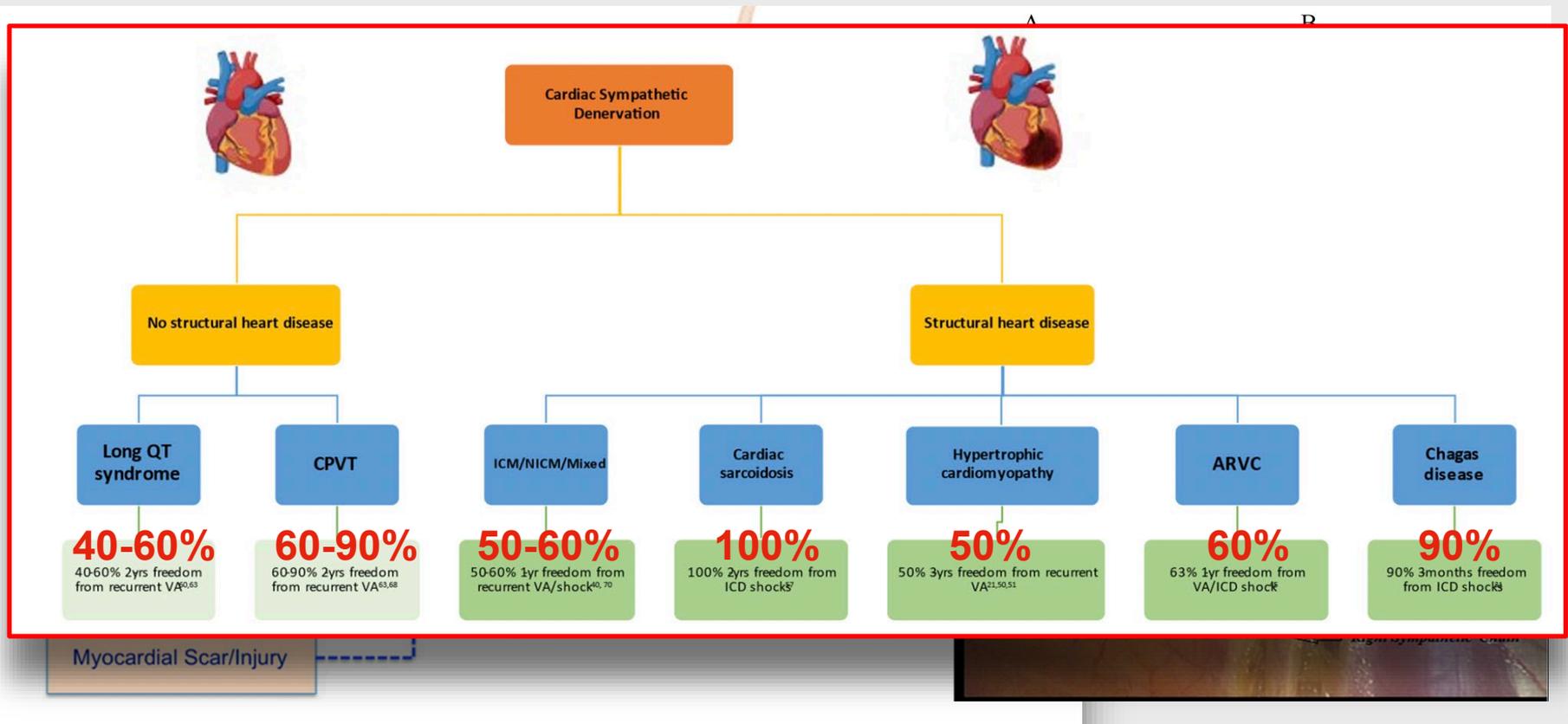
# Thoroskopische Sympathikus Denervierung

Kochav et al., *J of Interv Cardiac Electrophysiol* 2022;65:813-826



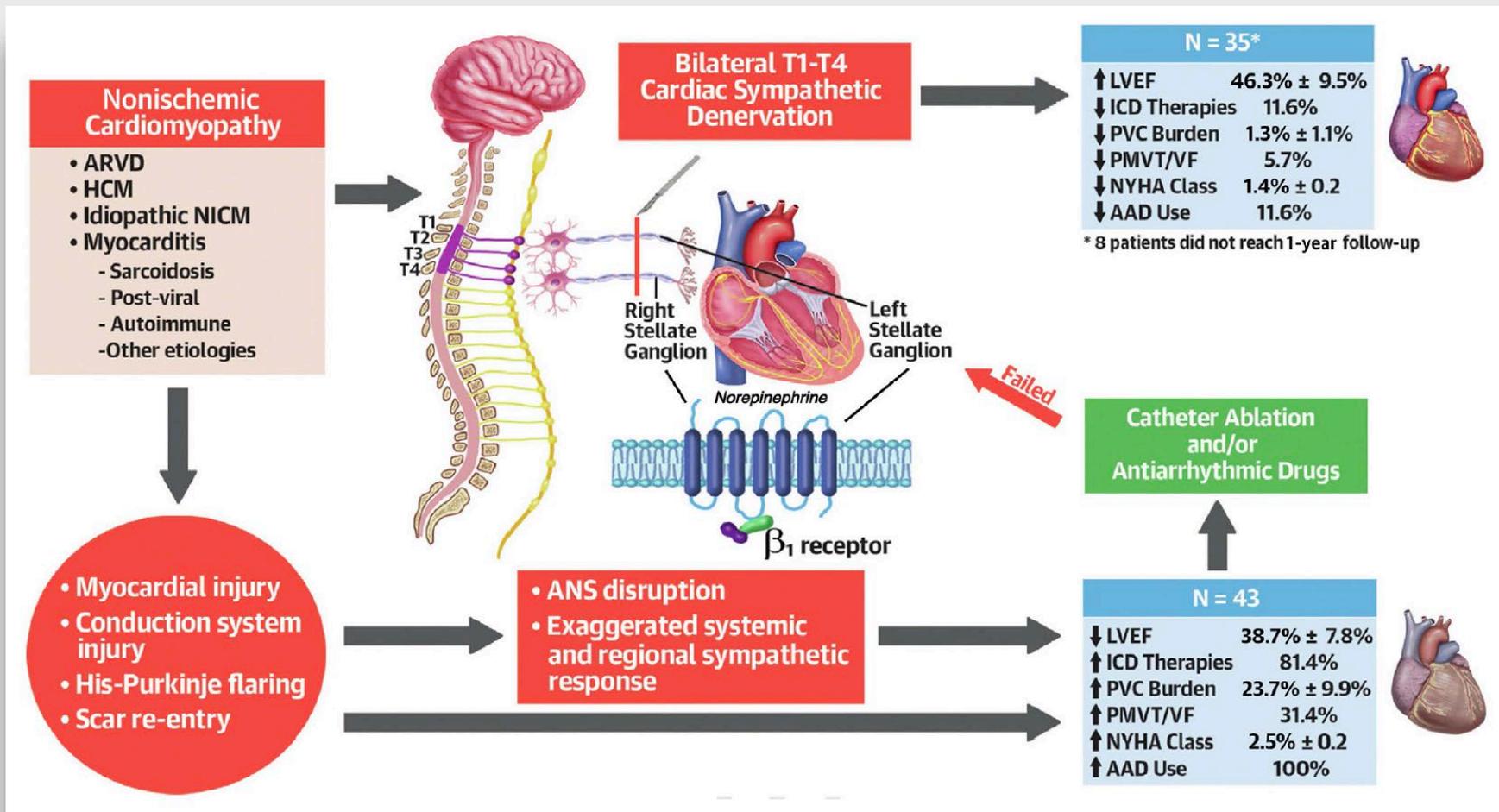
# Thoroskopische Sympathikus Denervierung

Kochav et al., *J of Interv Cardiac Electrophysiol* 2022;65:813-826



# Sympathikus Denervierung nicht ischämischer Kardiomyopathie und therapierefraktären VES

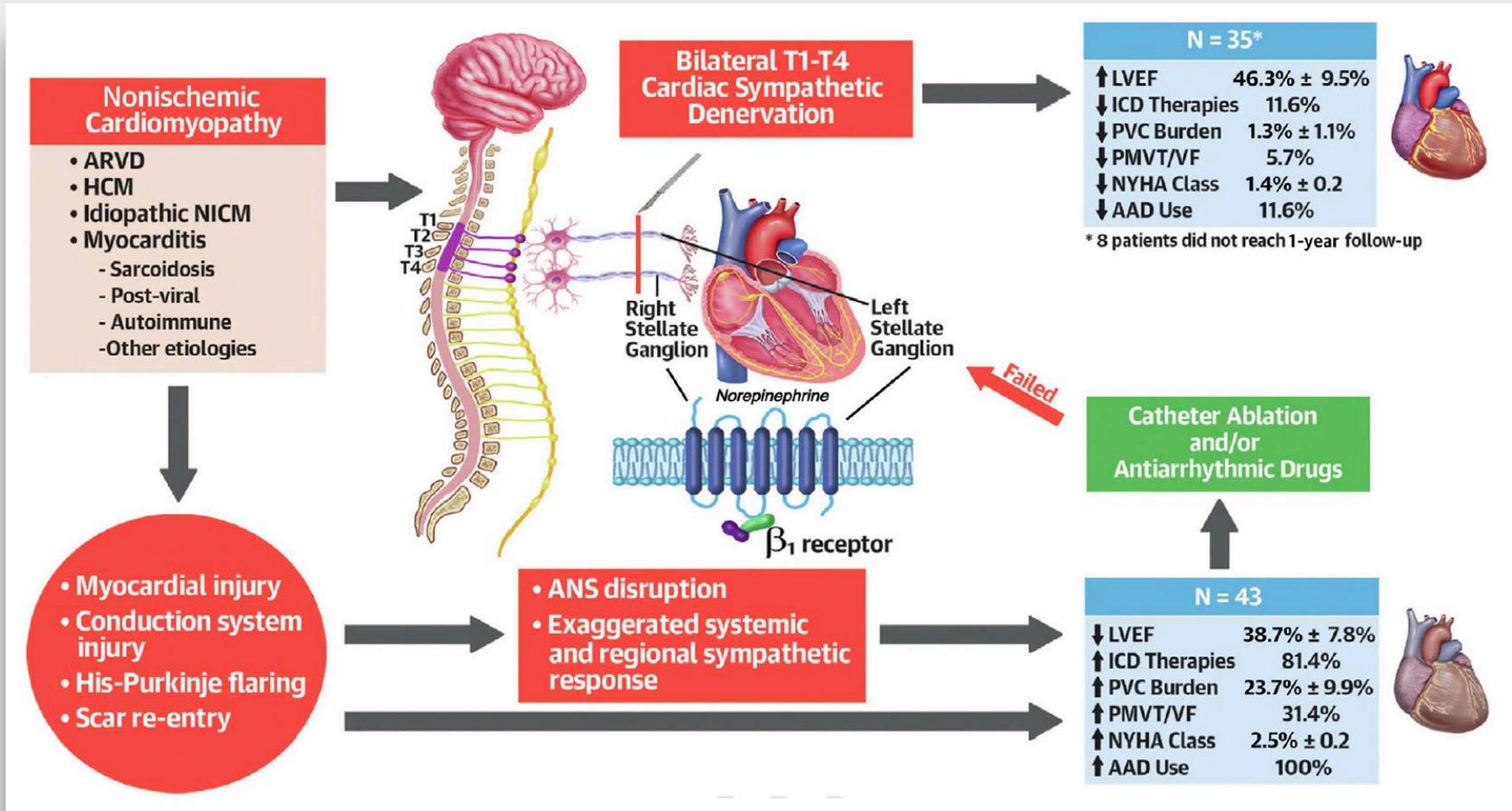
Ahmed et al., JACC CE 2024; 10:31-39



Bilateral Cardiac Sympathetic Denervation for Refractory Multifocal Premature Ventricular Contractions in Patients With Nonischemic Cardiomyopathy

# Denervierung nicht ischämischer und therapierefraktären VES

Ahmed et al., JACC CE 2024; 10:31-39



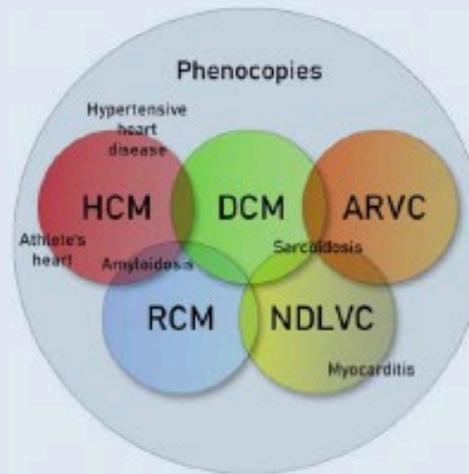
# Kardiomyopathien und Rhythmusstörungen

*Korthals & Eckardt; Herzschr Elektrophysiol 2023;34:311-323*

## Electrophysiologic findings or abnormalities in cardiomyopathies

### History

- Palpitations
- Arrhythmogenic syncope
- Exercise-related syncope
- Evidence for systemic (arrhythmogenic) disease
- Pacemaker/ICD implantation
- Fx for premature death (< 50yrs)
- Fx of ICD/pacemaker implant



### ECG

- Low/high QRS voltages
- Delayed conduction (e.g., atypical RBBB; QRS width ↑)
- T-Wave inversion (e.g., V1-V3 and terminal activation delay)
- AV block (I° - III°)
- Short PR/ pseudo pre-excitation
- Q waves/pseudo infarction

### Brady- or tachyarrhythmias

Symptomatic bradycardia, asystole  
 Conduction disease: high-grade AV block  
 Atrial fibrillation/flutter  
 Non-sustained Ventricular Arrhythmias/VEs  
 Sustained monomorphic VT  
 Sustained polymorphic VT/VF

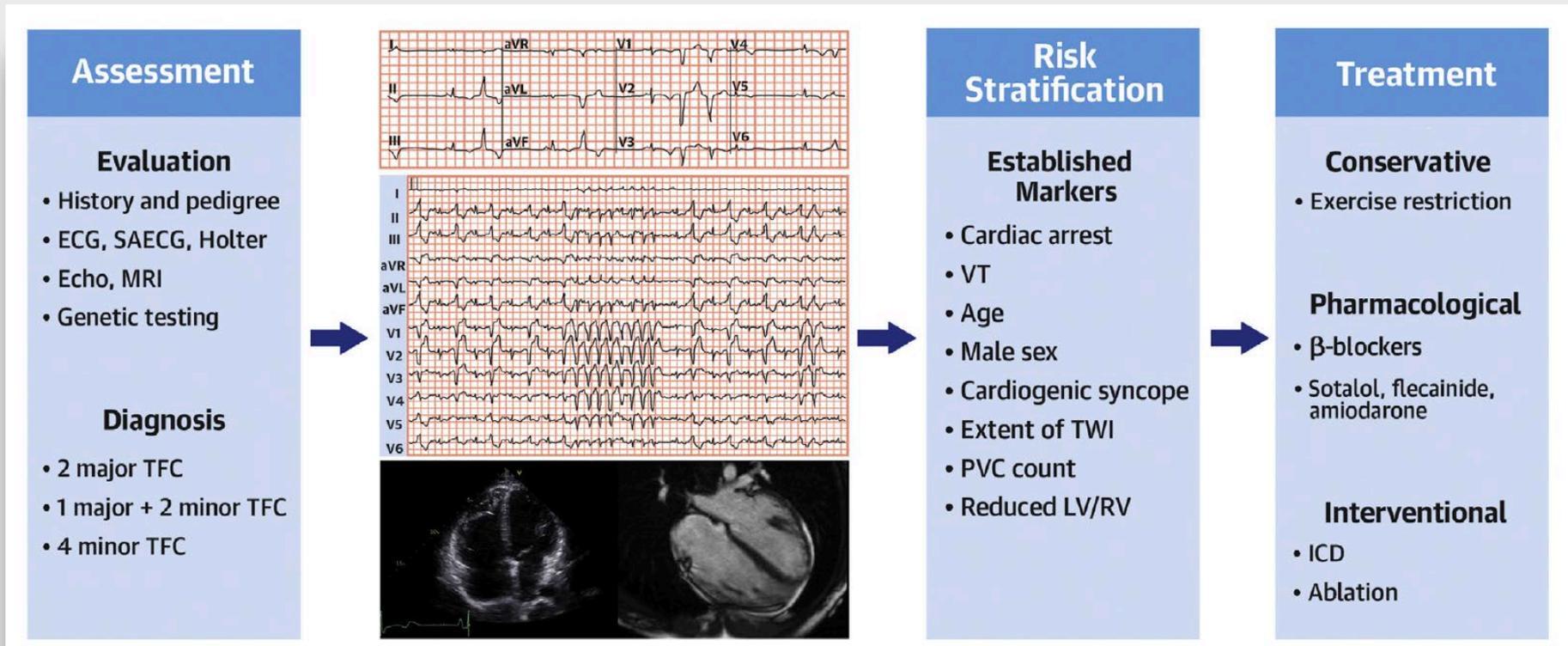


Pacemaker (discuss ICD and/or CRT)  
 Pacemaker (discuss ICD therapy)  
 OAC & rhythm vs. rate control  
 Risk stratification (discuss AAD/ablation)  
 ICD therapy (consider AAD/ablation)  
 ICD therapy (discuss AAD/ablation)

# **Arrhythmogene rechtsventrikuläre Kardiomyopathie (ARVC) und kardiale Sarkoidose**

# ARVC: State of the Art

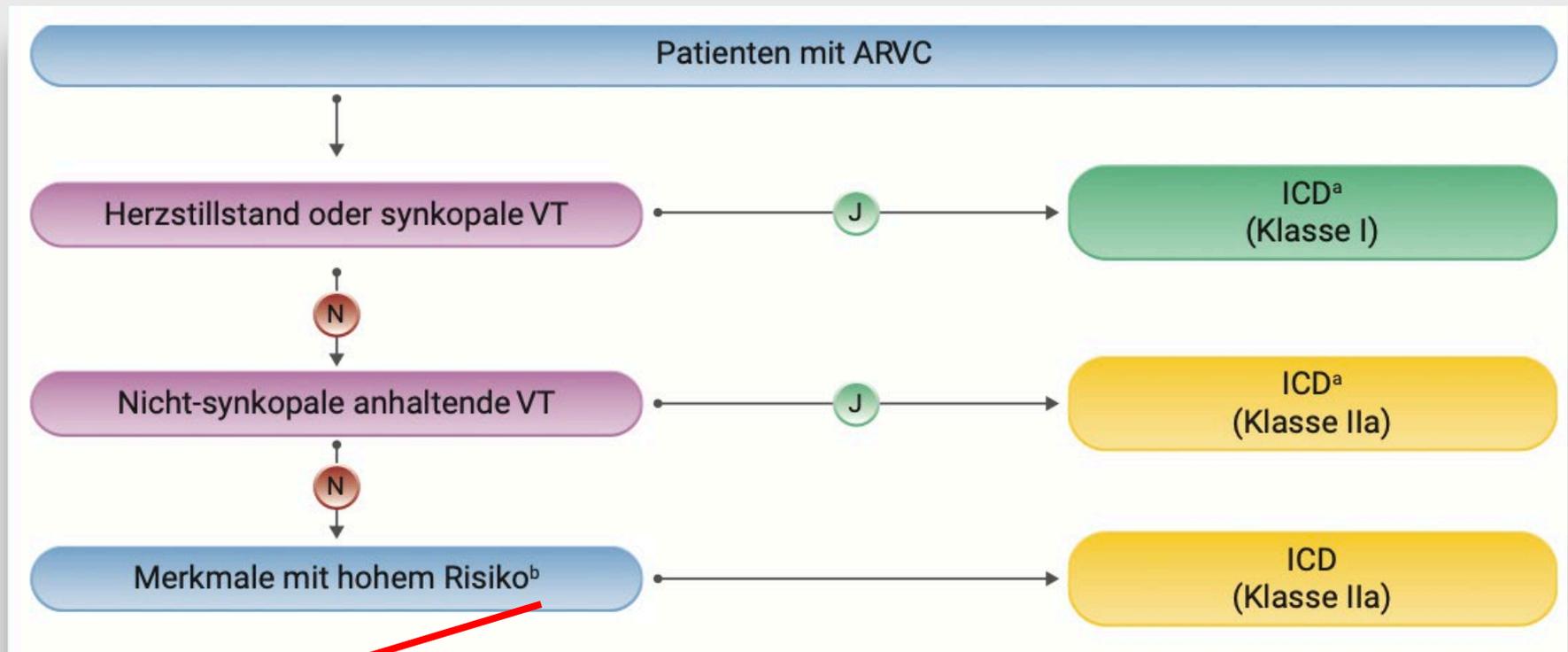
Kahn et al., J Am Coll Cardiol EP 2022;8:533-553



# ARVC: State of the Art

ESC Pocket GL 2022

Eckardt et al., Die Kardiologie 2023;17:27–38



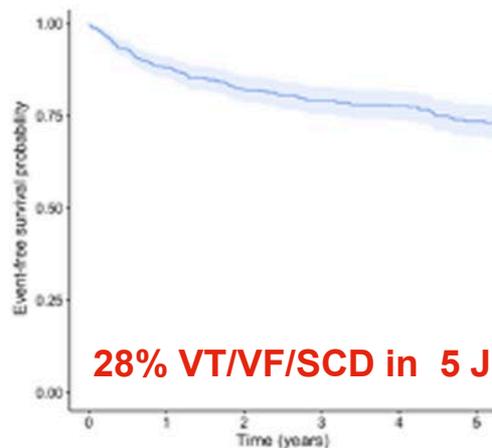
*Kardiale Synkope, NSVT, RVEF <40%, LVEF <45%, SMVT bei PES oder gemäß Risikokalkulator*

# ARVC – Risikostratifikation – Risiko Score (ESC GL IIa)

Cadrin-Tourigny et al., Eur Heart J 2022;43:e1-e9

## Prediction of sustained ventricular arrhythmia in ARVC (n=528)

5-year event-free survival (n = 528)  
Overall

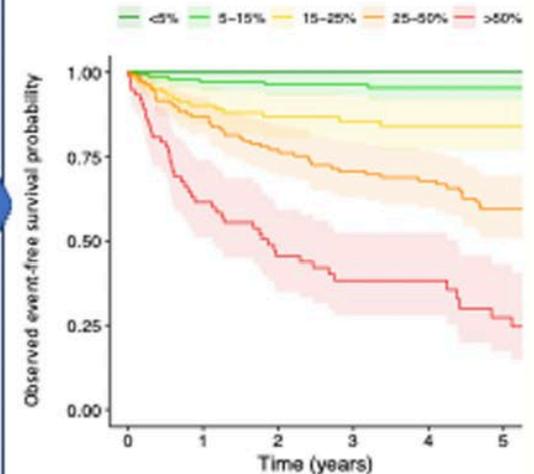


Model for 5-year risk prediction

Sex	x	0.49
Age	x	-0.022
Recent syncope	x	0.66
Non-sustained VT	x	0.81
Ln(24h PVC count)	x	0.17
Leads with T-wave inv.	x	0.11
RVEF	x	-0.025

----- +  
 $1 - 0.8396^{\exp(\dots)} = 5 \text{ year risk}$

5-year event-free survival  
Per predicted risk group



**Legend:** ARVC = arrhythmogenic right ventricular cardiomyopathy; VT = ventricular tachycardia; PVC = Premature ventricular complex; Inv, inversion; RVEF = right ventricular ejection fraction.

# ARVC – Risikostratifikation –Kammerstimulation

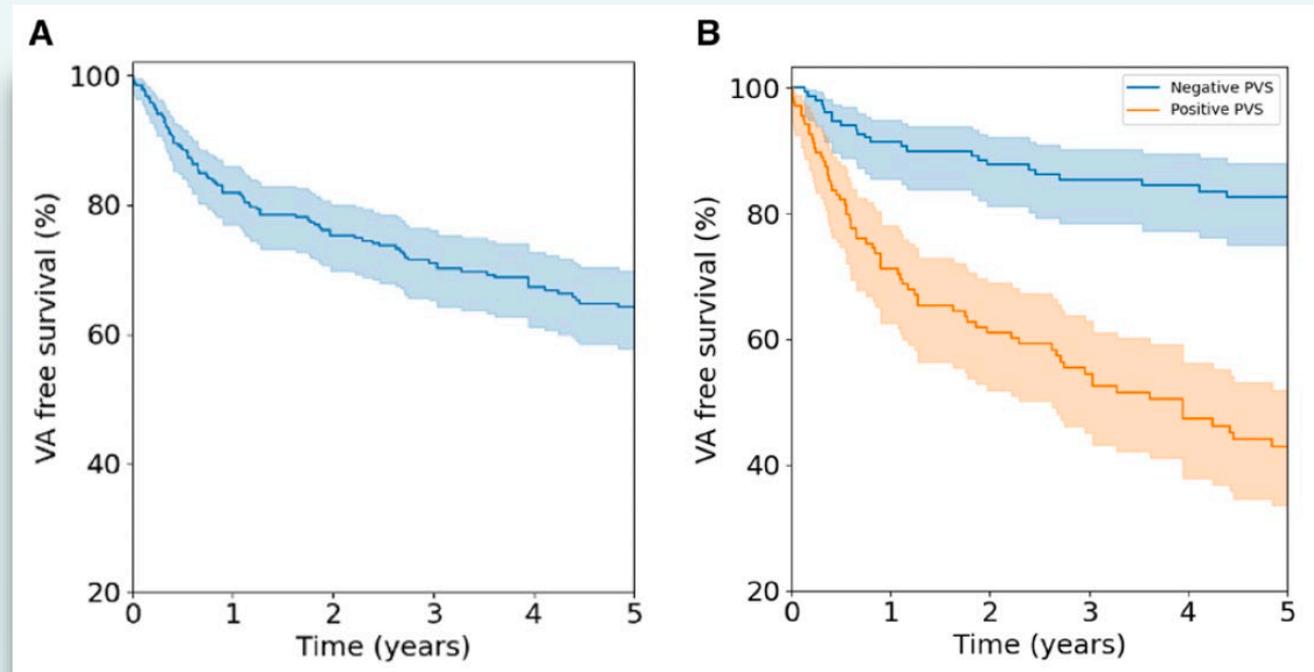
*Gasperetti et al., Circulation 2022;146:1434-1443*

288 ARVC Pat. (41.0±14.5 Jahre, 69% M) aus 6 internationalen Registern

Follow-up 5 Jahre

VT im Follow-up: **61% mit pos vs. 25% mit neg. PVS hatten VT (P=0,001)**

PVS mit  
signifikant  
verbessertes  
Aussagekraft  
des ARVC  
Risiko-  
Kalkulators.



Programmed Ventricular Stimulation as an Additional Primary Prevention Risk Stratification Tool in Arrhythmogenic Right Ventricular Cardiomyopathy: A Multinational Study

## Ammerstimulation

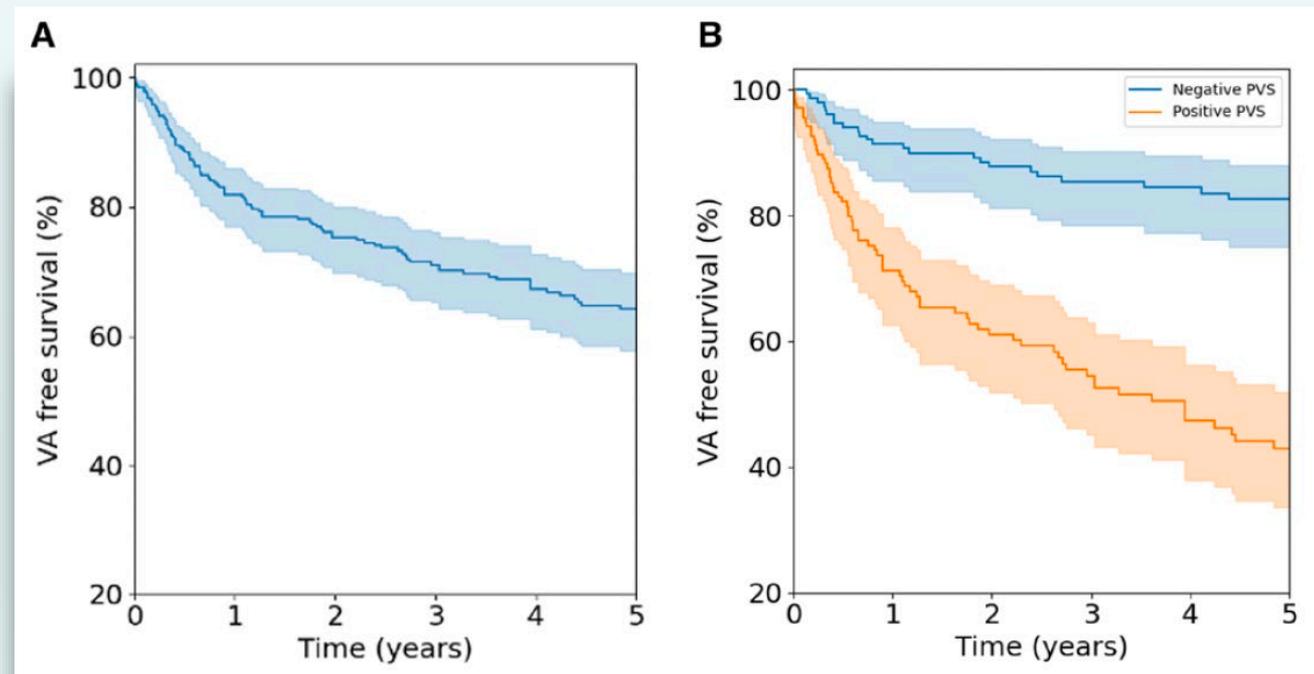
146:1434-1443

288 ARVC Pat. ( $41.0 \pm 14.5$  Jahre, 69% M) aus 6 internationalen Registern

Follow-up 5 Jahre

VT im Follow-up: **61% mit pos vs. 25% mit neg. PVS hatten VT** ( $P=0,001$ )

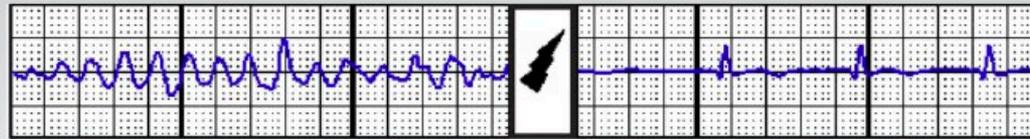
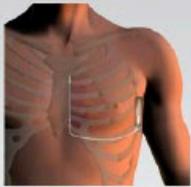
PVS mit  
signifikant  
verbesserter  
Aussagekraft  
des ARVC  
Risiko-  
Kalkulators.



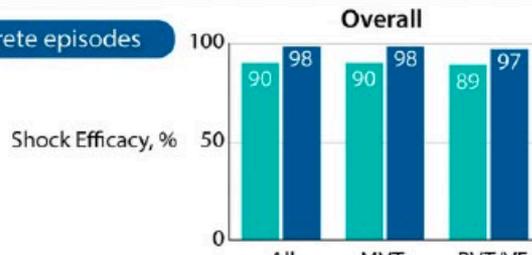
# S-ICD auch bei ARVC?

Lambiase et al., Eur Heart J 2022;43:2037-2050

## Maintenance of S-ICD Shock Efficacy over 5 years



### A. Discrete episodes



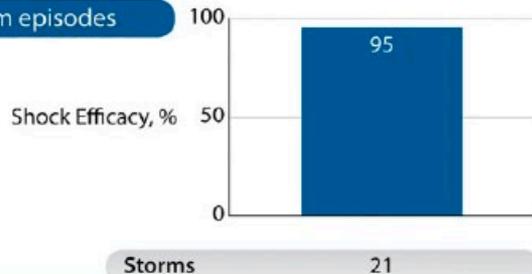
Number of episodes	310	161	149
Mean shocks per patient	1.1	1.1	1.1
Maximum shocks per patient	5	5	4

### Over time



Number of episodes	103	56	51	49	31	20
Mean shocks per patient	1.1	1.2	1.1	1.1	1.1	1.1
Maximum shocks per patient	5	5	2	2	2	2

### B. Storm episodes

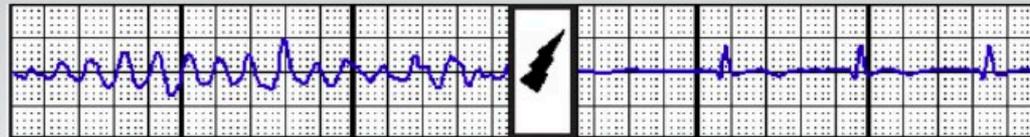
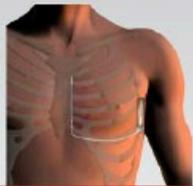


Number of episodes	7	8	4	0	2
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# S-ICD auch bei ARVC?

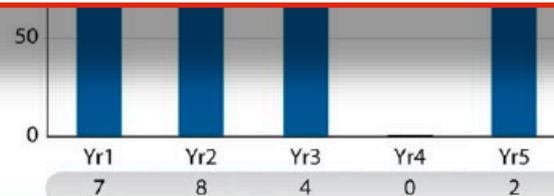
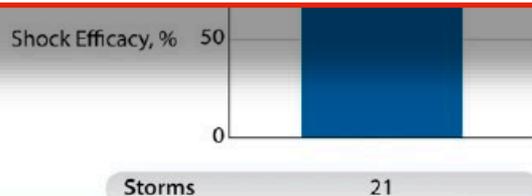
Lambiase et al., Eur Heart J 2022;43:2037-2050

Maintenance of S-ICD Shock Efficacy over 5 years



## B. Appropriate Therapy for MVT

ARVC vs no ARVC	7.10 (2.68-18.78)	
Cardiac Arrest vs no Cardiac Arrest	2.48 (1.49-4.15)	
Appropriate Shock (AS) Year 1 vs no AS Year 1	3.09 (1.57-6.09)	
CHF vs no CHF	1.99 (1.20-3.30)	
Ischemic vs non-Ischemic	2.00 (1.12-3.58)	
Self-terminating Appropriate Episode (STAE) Year 1 vs no STAE Year 1	2.39 (1.14-5.02)	
NYHA Class III or IV vs Class I or II	0.24 (0.07-0.82)	
Inappropriate Shock (IAS) Year 1 vs no IAS Year 1	1.85 (0.95-3.60)	
Shock Zone (per 10 bpm)*	0.87 (0.74-1.02)	
NICM/DCM vs no NICM/DCM	1.80 (0.90-3.61)	
Valve Surgery vs no Valve Surgery	0.47 (0.16-1.34)	
Conversion Testing (CT) within 30 Days vs no CT within 30 days	0.63 (0.27-1.48)	

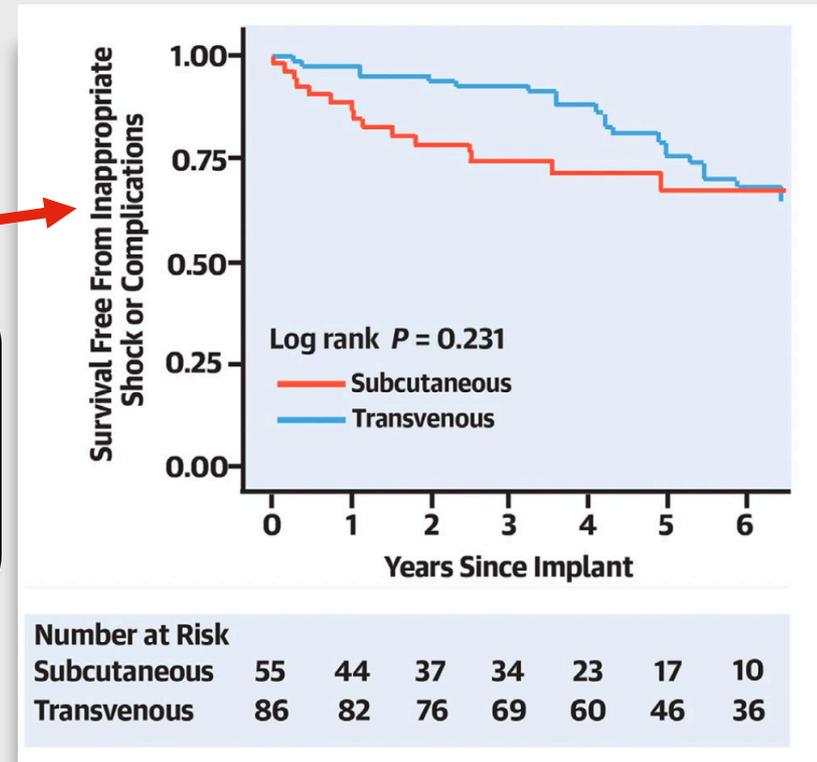


# S-ICD auch bei ARVC?

Wang W et al., JACC EP 2023;9:394-402

- Match control study (57 vs 88) (35 Jahre; 35% Sekundärprophylaxe)
- Schocks/Komplikationen und Lebensqualität

- S-ICD mehr inadäquate Schocks
  - 23% vs. 10%
- S-ICD weniger Device Komplikationen
  - 4% vs. 14%



# Subcutaneous and Transvenous Defibrillators in Arrhythmogenic Right Ventricular Cardiomyopathy

A Comparison of Clinical and Quality-of-Life Outcomes

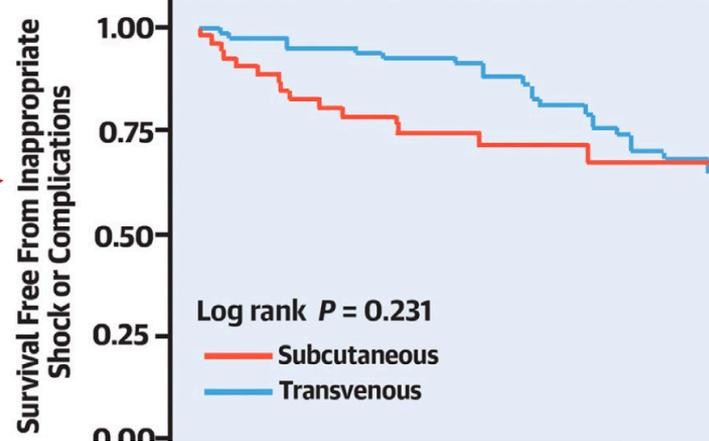
# ei ARVC?

2023;9:394-402

- Match control study (57 vs 88) (35 Jahre; 35% Sekundärprophylaxe)
- Schocks/Komplikationen und Lebensqualität

## S-ICD mehr inadäquate Schocks

- 23% vs. 10%



## Quality of Life Outcomes at 1-Year Follow-Up

	Subcutaneous n = 19	Transvenous n = 42	P Value
Range of motion being affected by the device*	2 (12%)	0 (0%)	0.021
Physical attractiveness affected by the device*	6 (38%)	1 (2%)	<0.001

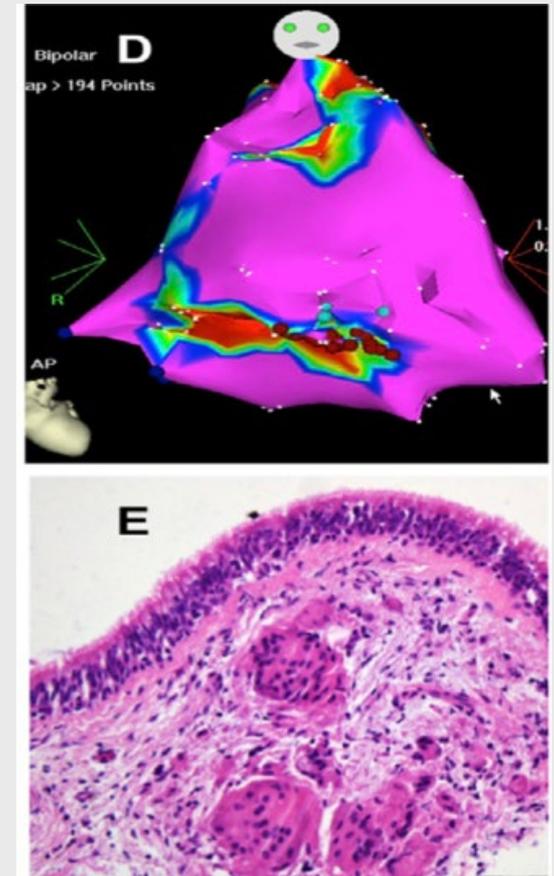
6

10  
36

# Sarkoidose: State of the Art

*Rosenthal et al., J Am Heart Assoc 2021;10:e017692*

- Multisystemerkrankung mit 2 Phasen (Entzündung und Fibrose)
- Ca. 5% klinisch manifeste kardiale Beteiligung
- Trias aus Leitungsstörungen, VT/VF und Herzinsuffizienz
- Zunehmende Diagnose in den vergangenen Jahren (wachsende Bedeutung der kardialen Bildgebung, MRT, FDG PET)
- Ca. 15% primär rhythmogene Manifestation mit VT/VF oder plötzlichem Herztod
- Frühe Diagnose aufgrund der Option einer immunsuppressiven Therapie entscheidend



*Willy et al., J Cardiol (2021) 77:154-159*

# Kardiale Sarkoidose – Manifestationen

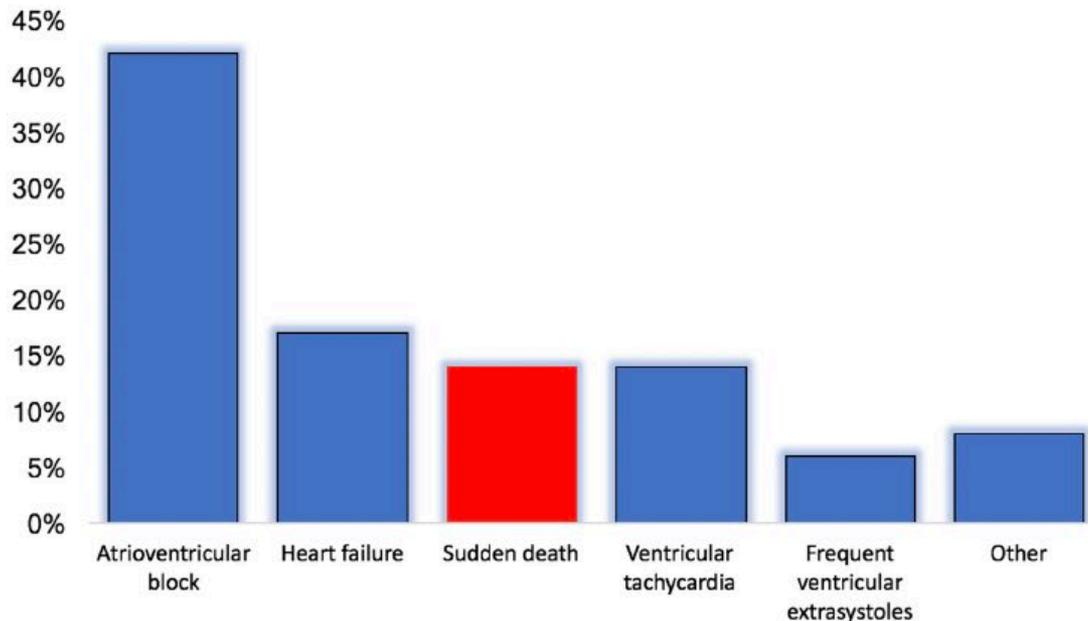
*Lehtonen et al., Eur Heart J 2023;44:1494-1510*

*Ekström et al. Eur Heart J 2019;40:3121-3128*

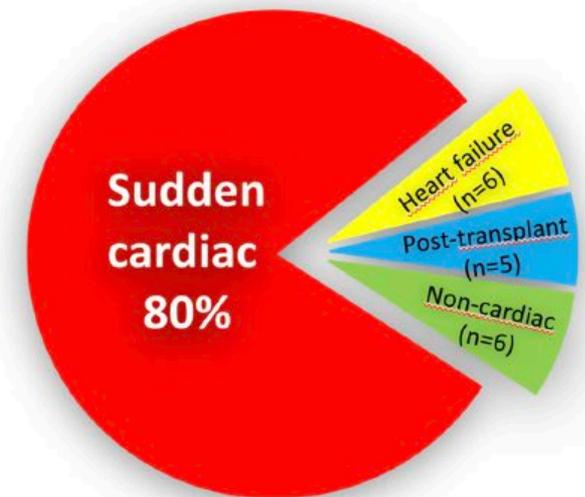
## Sudden death in cardiac sarcoidosis

Analysis of a nationwide 18-year case series from Finland (n=351)

### Main presenting manifestations



### Mode of deaths (n=84)



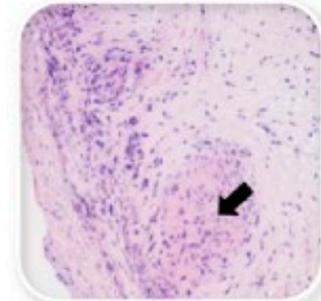
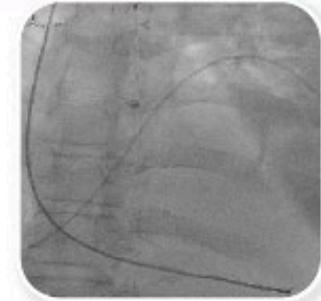
### Pathogenesis

- Unknown environmental triggers
- Genetic predisposition
- Granulomas → fibrotic scarring

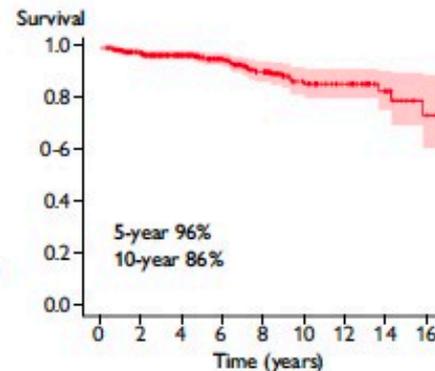
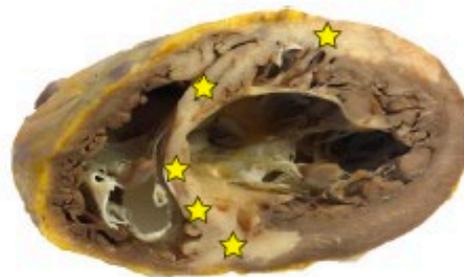
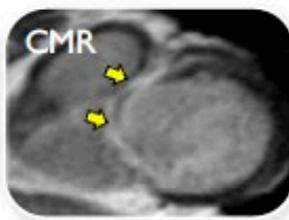
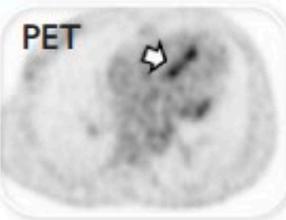
### Main manifestations

- Subclinical cardiomyopathy
- Atrio-ventricular block
- Ventricular tachycardia (VT)
- Heart failure

### Diagnostic biopsy



### Diagnostic imaging



### Therapy

- Tiered immunosuppression
- Treatment of arrhythmias and heart failure
- Consideration of an ICD to prevent SCD

### Factors predictive of worse outcome

- Extent of myocardial involvement (several indices)
- Presentation with VT or heart failure
- De novo or clinically isolated cardiac involvement
- Definite vs. probable diagnosis

45%  
40%  
35%  
30%  
25%  
20%  
15%  
10%  
5%  
0%

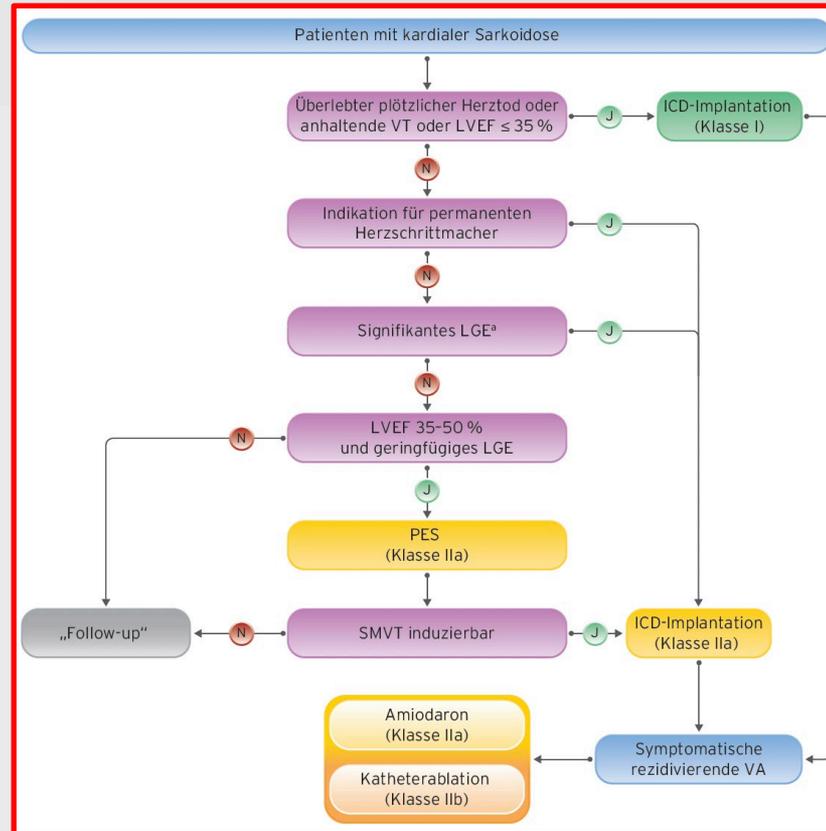
block

tachycardia

ventricular  
extrasystoles

# ESC-Leitlinie: VT und SCD

Zeppenfeld et al., *Eur Heart J* 2022; 43:3997-4126

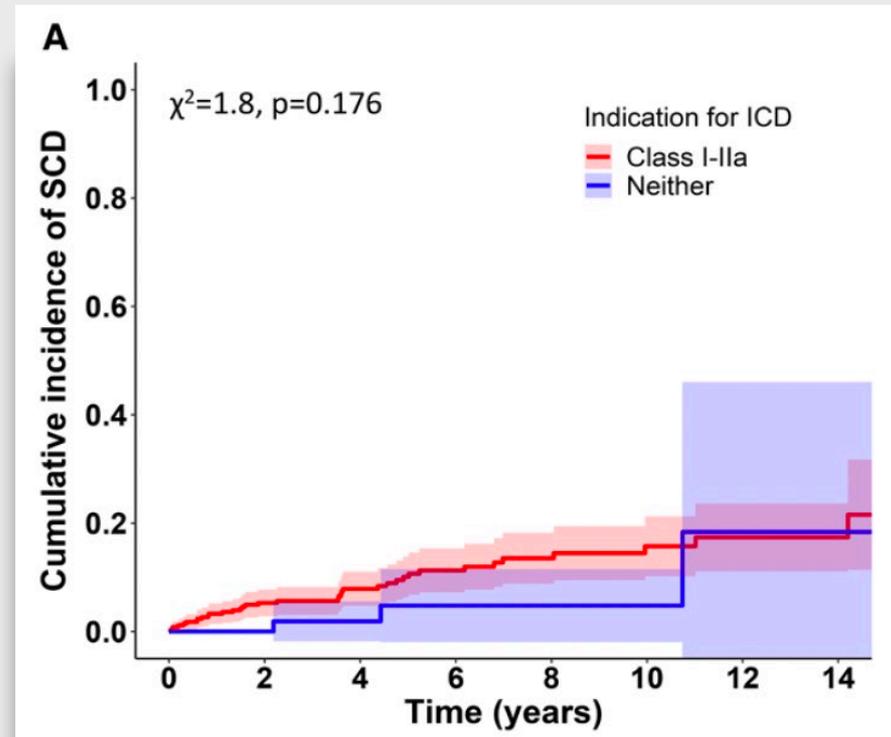


# Schwächen der Risikostratifikation

Nordenswan HK et al., *Circulation* 2022;146:964-975

- 398 Pat. mit kardialer Sarkoidose
- (40% histologisch gesichert)
- Follow-up 5 Jahre
- 10% „fatal or aborted) SCD

Guidelines	I / IIa	No I / IIa
5yr SCD rate	11%	5%
Definitive CS	14%	<b>6%</b>
Probable CS	8%	3%



In aktuellen LL werden Pat. mit kardialer Sarkoidose (CS) und (wirklich) niedrigem SCD Risiko nicht gut abgebildet.

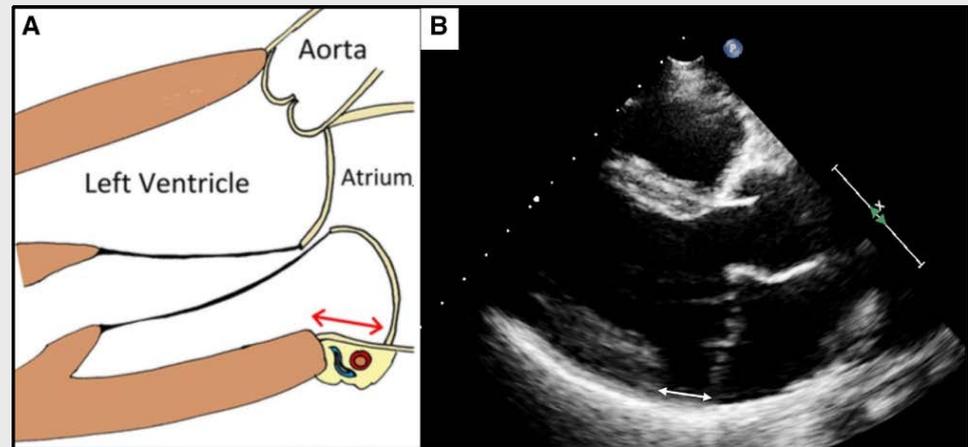
# Mitralklappenprolaps - Syndrom

# State of the Art

## Mitralklappen Prolaps Syndrom

- Mitralklappenprolaps **häufig** und überwiegend Zufallsbefund; geschätzte Häufigkeit bei **2-3%**
- **Definition:** Vorwölbung anteriores und/oder posteriores Segel in den Vorhof um  $\geq 2$  mm während der Systole in der parasternalen oder apikalen lange Achse
- PubMed 1953-2023 **ca. 8000 Treffer zum MVP**, davon etwa **420 Kombination plötzlicher Herztod und MVP**
- **Möglicher Marker** für einen plötzlichen Herztod (PHT)

### MAD = Mitral Annular Dysjunction



Dejgaard et al., *J Am Coll Cardiol.* 2018;72(14):1600-1609  
Basso et al. *Circulation.* 2019;140:952-964.

# Mitralklappen Prolaps Syndrom

*Nishimura et al., NEJM 1985; 313:1305-1309*

Volume 313

NOVEMBER 21, 1985

Number 21

## ECHOCARDIOGRAPHICALLY DOCUMENTED MITRAL-VALVE PROLAPSE

### Long-Term Follow-up of 237 Patients

RICK A. NISHIMURA, M.D., MICHAEL D. McGOON, M.D., CLARENCE SHUB, M.D.,  
FLETCHER A. MILLER, JR., M.D., DUANE M. ILSTRUP, M.S., AND A. JAMIL TAJIK, M.D.

**Abstract** We determined the long-term prognosis for patients with mitral-valve prolapse documented by echocardiography by following 237 minimally symptomatic or asymptomatic patients for a mean of 6.2 years (range, 1 to 10.4). The actuarial eight-year probability of survival was 88 per cent, which is not significantly different from that for a matched control population. An initial left ventricular diastolic dimension exceeding 60 mm was the best echocardiographic predictor of the subsequent need for mitral-valve replacement (17 patients). Of the 97 patients with redundant mitral-

valve leaflets identified echocardiographically, 10 (10.3 per cent) had sudden death, infective endocarditis, or a cerebral embolic event; in contrast, of the 140 patients with nonredundant valves, only 1 (0.7 per cent) had such complications ( $P < 0.001$ ). Most patients with echocardiographic evidence of mitral-valve prolapse have a benign course, but subsets at high risk for the development of progressive mitral regurgitation, sudden death, cerebral embolic events, or infective endocarditis can be identified by echocardiography. (N Engl J Med 1985; 313:1305-9.)

**M**ITRAL-valve prolapse is a common entity<sup>1</sup> that has been implicated in several cardiac syndromes, ranging from the nonspecific symptoms of atypical chest pain, anxiety, and palpitations<sup>2</sup> to more devastating manifestations such as cerebral embolic

leaflet redundancy. The criterion for unequivocal mitral-valve prolapse was late or holosystolic bowing of the mitral-valve leaflet 3 mm or more below the C-D line (the line of mitral-leaflet coaptation during systole) (range, 3 to 22 mm; mean, 7 mm). Patients under 10 years old or over 70 were excluded, as were patients with associated congenital anomalies or other valvular diseases. Patients

# Mitralklappen Prolaps Syndrom

*Nishimura et al., NEJM 1985; 313:1305-1309*

Volume 313

NOVEMBER 21, 1985

Number 21

## ECHOCARDIOGRAPHICALLY DOCUMENTED MITRAL-VALVE PROLAPSE

RICK A. N  
FLETCHER A.

**Abstract** We determined patients with mitral-valve prolapse by following 237 mitral-valve prolapse patients for a mean 8.5 years. The actuarial eight-year probability of survival was 93 percent, which is not significantly different from the matched control population. The left ventricular dimension exceeding 60 mm was a significant predictor of the subsequent need for mitral valve replacement (17 patients). Of the 9

**M**ITRAL-valve prolapse has been implicated in a variety of syndromes, ranging from the atypical chest pain, anxiety, and other noncardiac manifestations to the devastating manifestations of

E — NISHIMURA ET AL.

1307

### Predictors of Complications

#### *Progressive Mitral Regurgitation Requiring Operation*

The left ventricular diastolic dimension, left ventricular systolic dimension, and left atrial size were all significant univariate echocardiographic predictors of progressive mitral regurgitation requiring mitral-

# Mitralklappen Prolaps Syndrom

*Nishimura et al., NEJM 1985; 313:1305-1309*

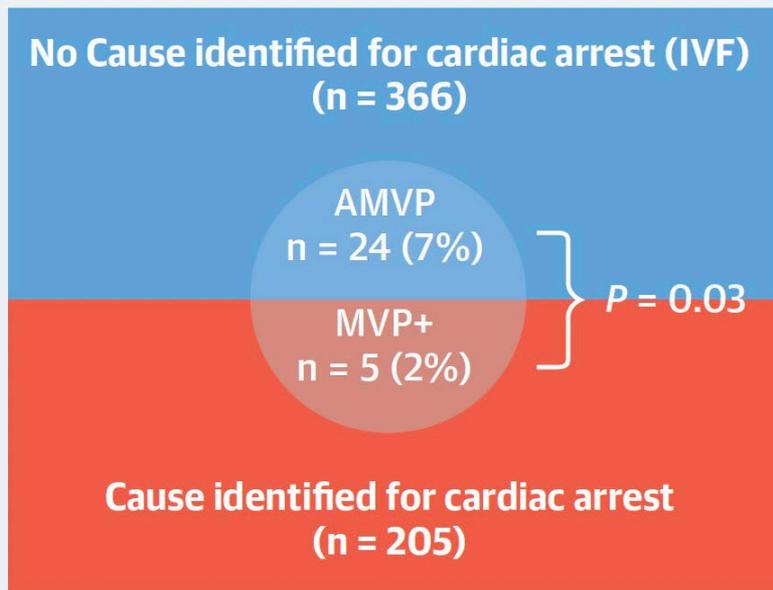
*Alqarawi et al., JACC CE 2023; 9:2494-2503*

Volume 313

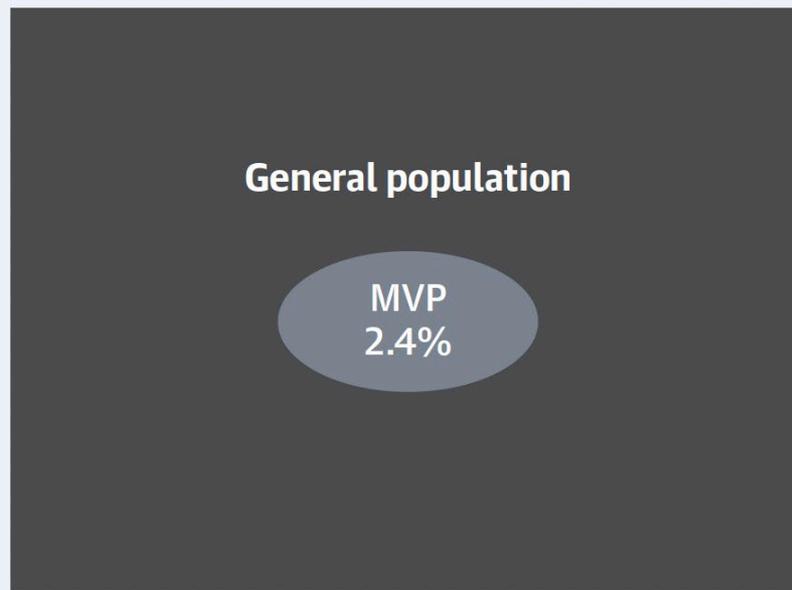
NOVEMBER 21, 1985

Number 21

## UCA Population:



## General Population:



IVF has been implicated in various arrhythmias, ranging from the benign to the life-threatening. In some cases, it is associated with atypical chest pain, anxiety, and other symptoms. In severe cases, it can lead to devastating manifestations such as sudden cardiac death.

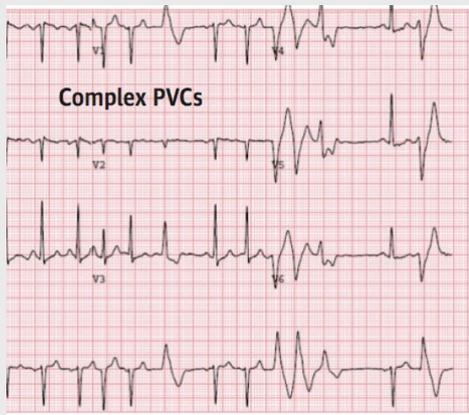
all significant univariate echocardiographic predictors of progressive mitral regurgitation requiring mitral-

# Mitralklappen Prolaps Syndrom

Muthukumar et al., JAMA Cardiol 2020;5:1053-1061

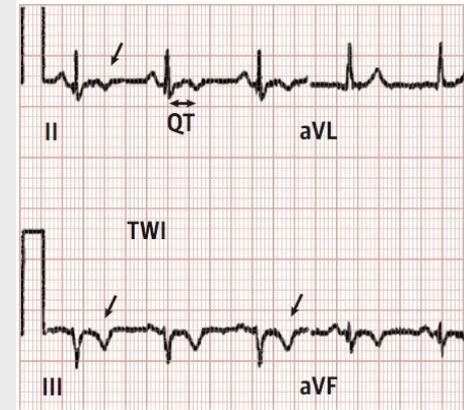
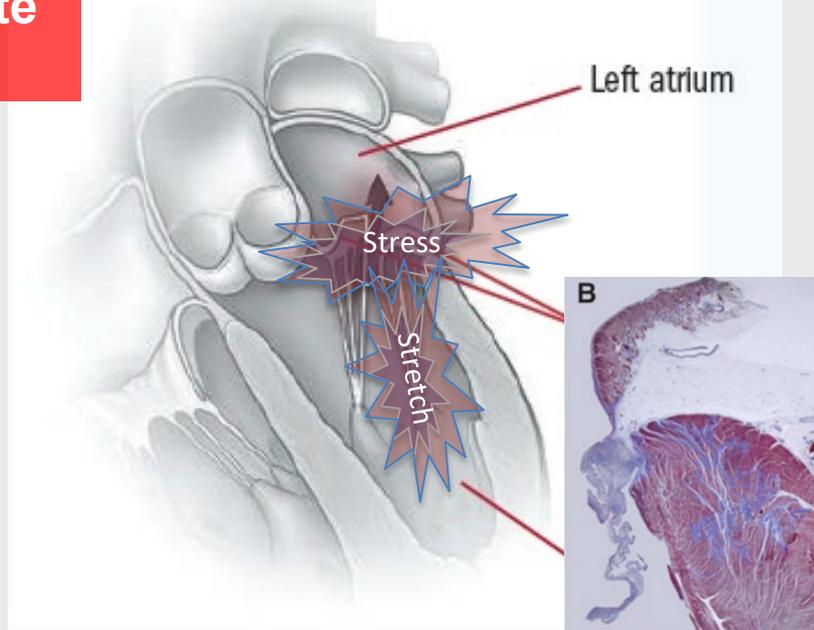
## Second Hit Theory

Mechanisch getriggerte  
Extrasystolen



Trigger

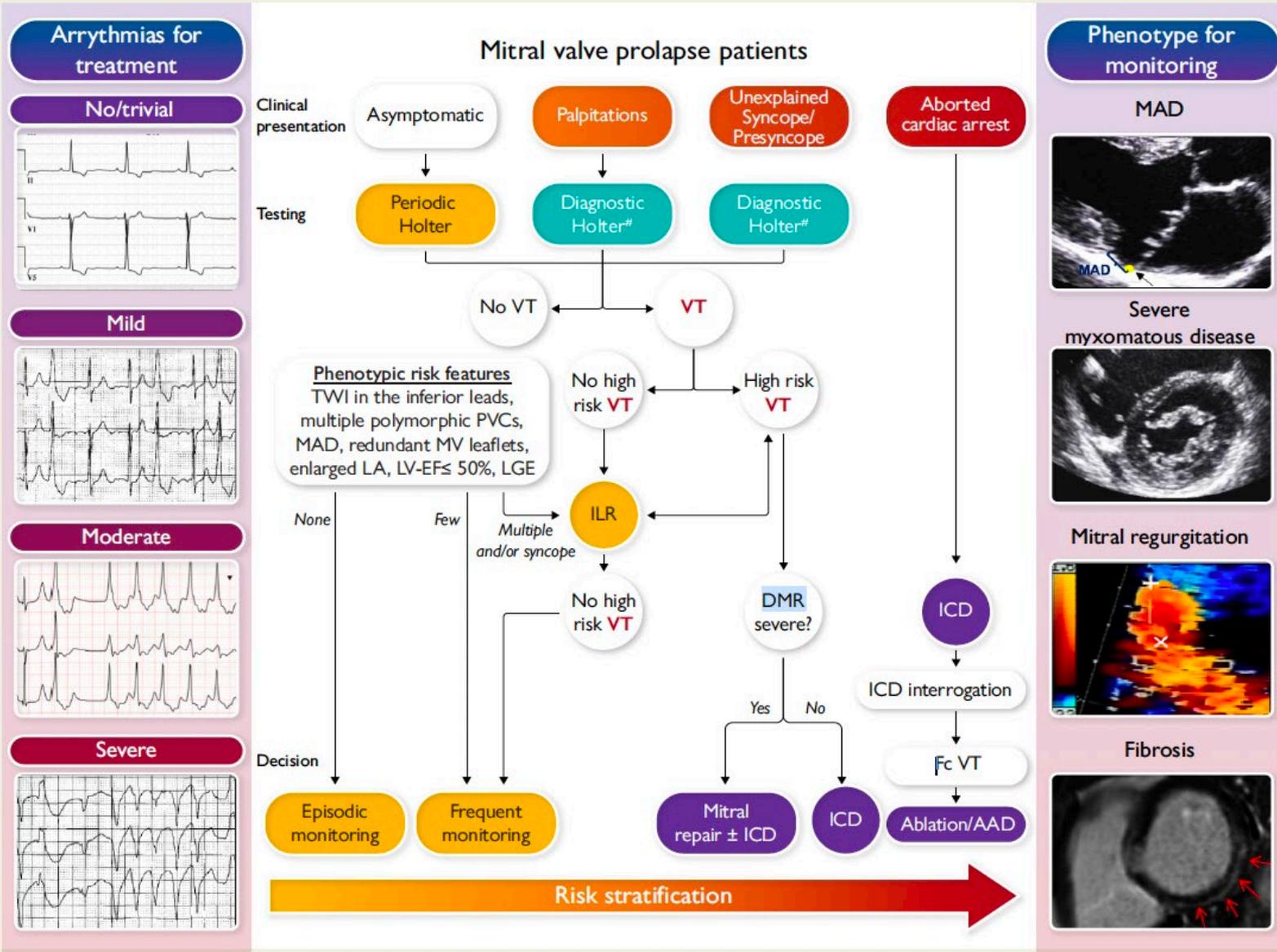
Repolarisationsstörungen



Stretch- und  
kontaktions-  
induzierte  
Fibrose

Substrat

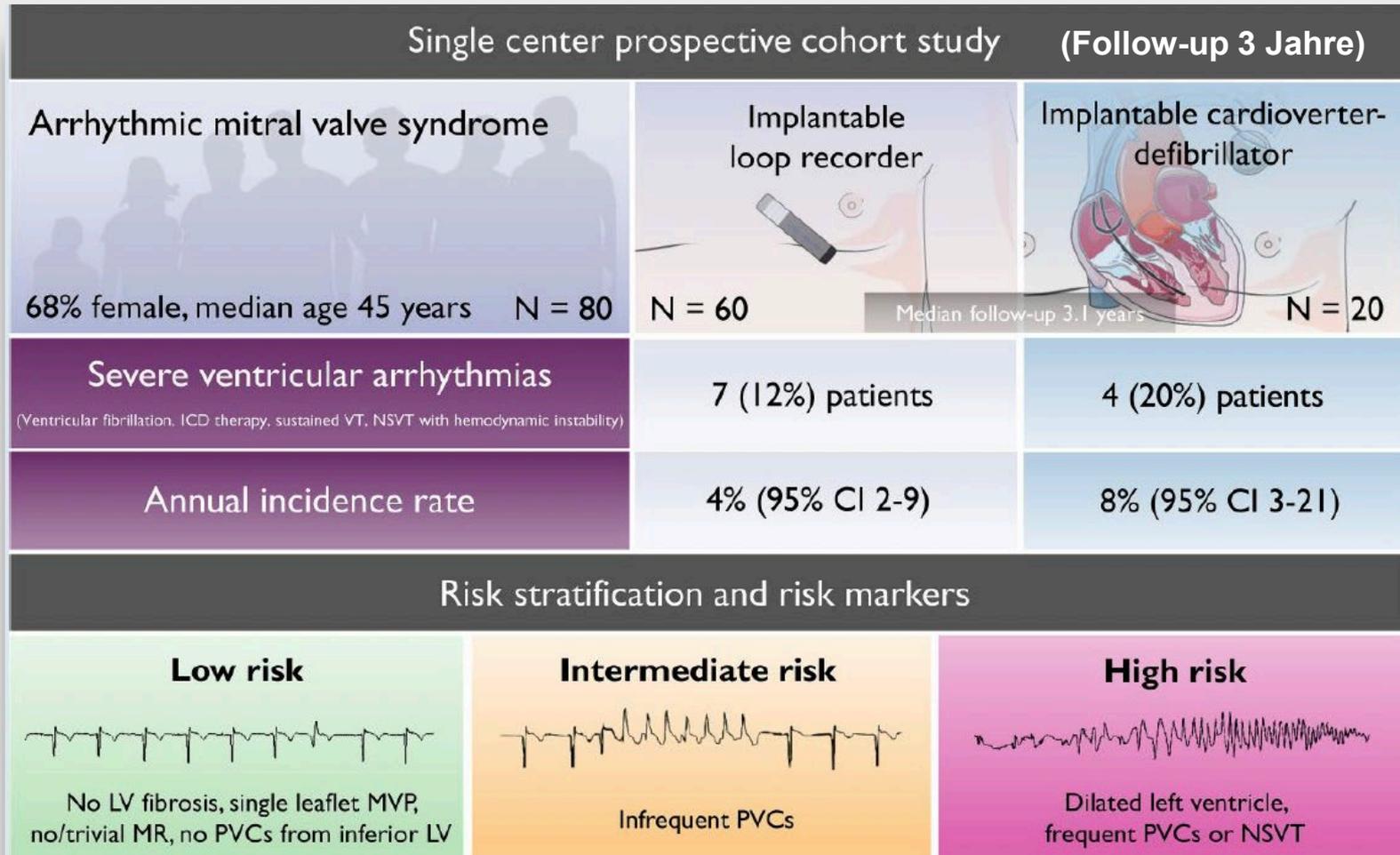
# Mitral valve prolapse syndrome



Essayagh et al., Euro Heart J (2023) 44, 3121–3135

# Mitralklappen Prolaps Syndrom

*Aabel et al., Europace 2023;25:506-516*



# Ionenkanal Erkrankungen

# ESC-Leitlinie: Aufwertung der Genetik

Zeppenfeld et al., *Eur Heart J* 2022;43:3997-4126

Eckardt et al., *Die Kardiologie* 2023;17:27–38

**2022 NEU!**

## Klasse I Empfehlungen zur genetischen Diagnostik

### Kanalerkrankungen

Empfehlungen für primär elektrische Erkrankungen
Klinisch diagnostiziertes Langes-QT-Syndrom
Vermutetes Andersen-Tawil-Syndrom
Brugada-Syndrom
Vermutetes oder klinisch diagnostizierte <u>katecholaminerge polymorphe ventrikuläre Tachykardie</u>
Kurzes-QT-Syndrom

### Strukturelle Herzerkrankungen

Empfehlungen für spezifische strukturelle Herzerkrankungen
Dilatative Kardiomyopathie/ hypokinetische nicht-dilatative Kardiomyopathie und <ul style="list-style-type: none"><li>- AV-Überleitungsstörungen vor dem 50. Lebensjahr</li><li>- Positive Familienanamnese für dilatative Kardiomyopathie/ hypokinetische nicht-dilatative Kardiomyopathie bei einem Verwandten ersten Grades (vor dem 50. Lebensjahr)</li></ul>
Vermutete oder bestätigte arrhythmogene rechtsventrikuläre Kardiomyopathie
Hypertrophe Kardiomyopathie

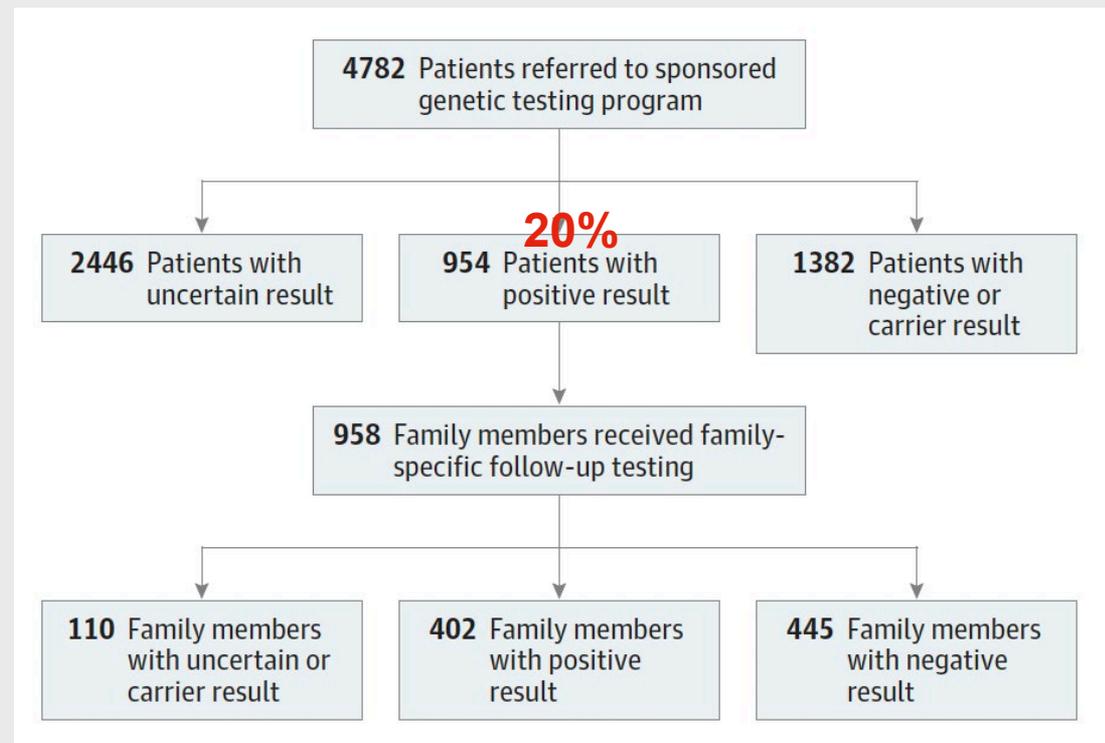
© L.Eckardt

# Assessment of the Diagnostic Yield of Combined Cardiomyopathy and Arrhythmia Genetic Testing

*Dellefave-Castillo et al., JAMA Cardiol 2022;9:966-974*

Retrospektive Analyse von 4782 Pat. (41 Jahre); single center, ein Jahr

- 50% mit Nachweis von genetischen Varianten unklarer Signifikanz.
- Eindeutig pos. Ergebnis 20%
- 2/3 davon Einfluss auf Entscheidungen
- Kaskadenuntersuchung identifizierte 42% pos Familienangehörige

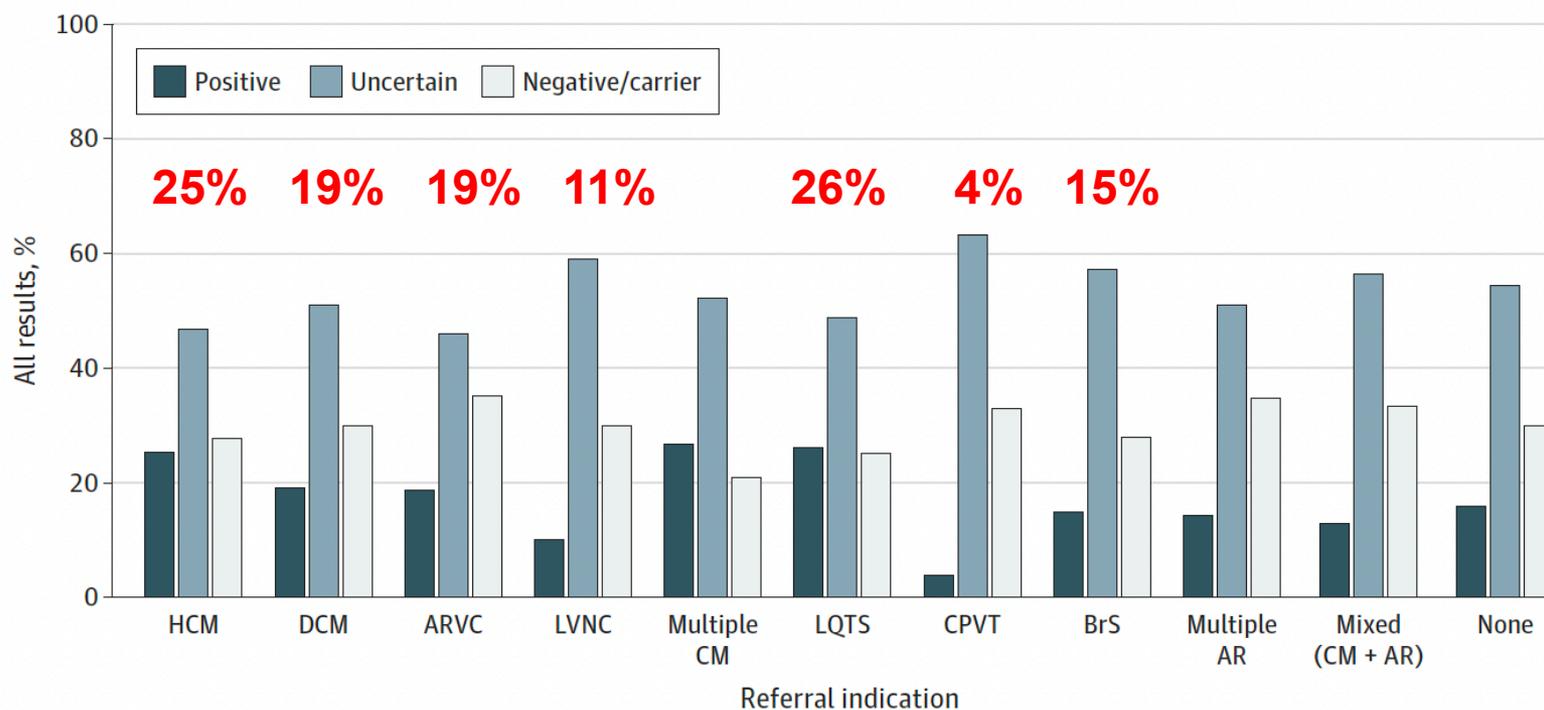


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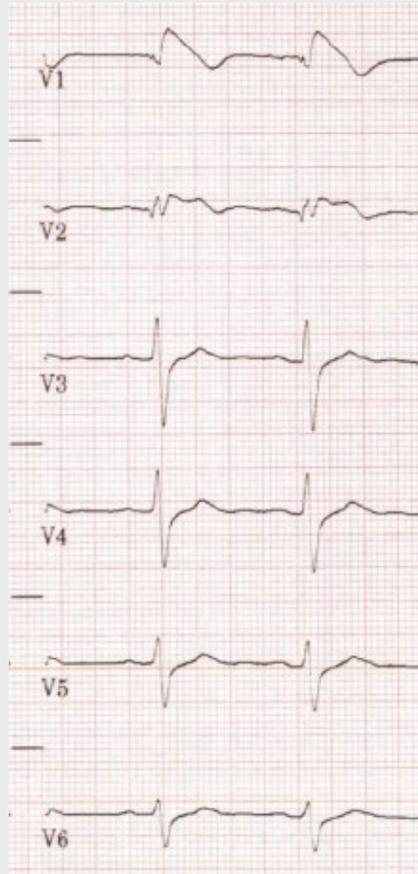
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# Brugada Syndrom

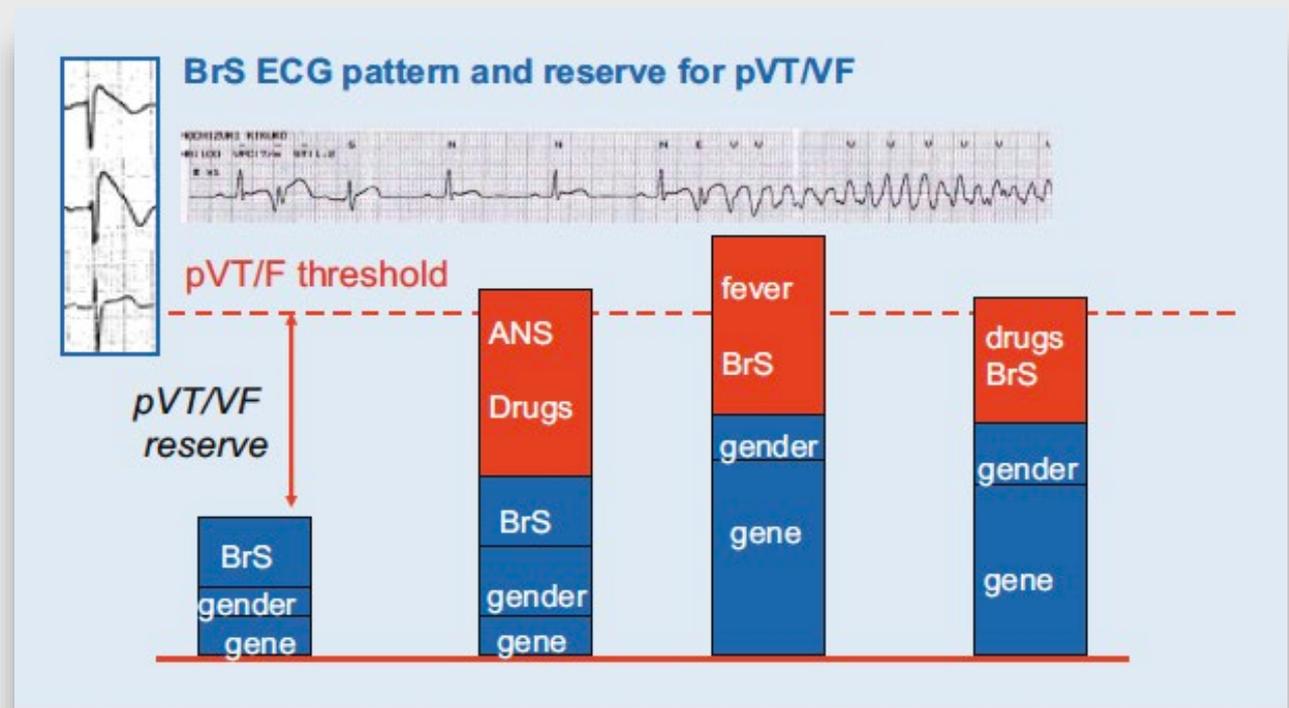
Brugada et al., JACC 2018; 72:1046-1059

Eckardt & Veltmann, Herzschr Elektrophysiol 2023; doi: 10.1007/s00399-023-00983-y



25mm/s

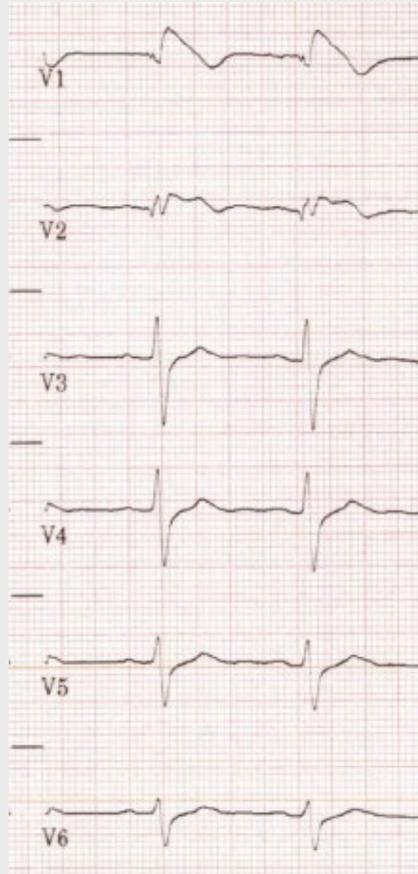
- ST-Hebung mit Typ I Morphologie  $\geq 2\text{mm}$  in  $\geq 1$  Abl. V1-V3
- Verantwortlich für 5-10% aller SCD; 20% der SCD ohne str. HE



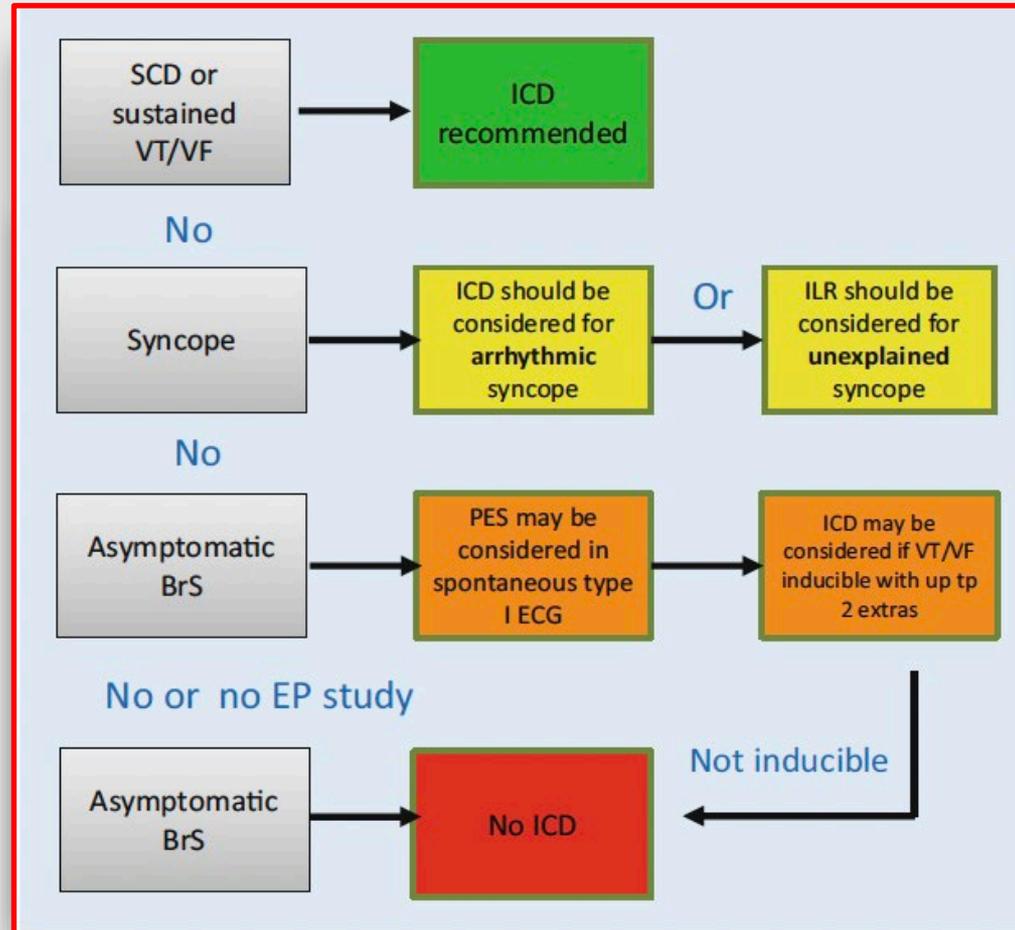
# Brugada Syndrom

Brugada et al., JACC 2018; 72:1046-1059

Eckardt & Veltmann, Herzschr Elektrophysiol 2023; doi: 10.1007/s00399-023-00983-y



25mm/s



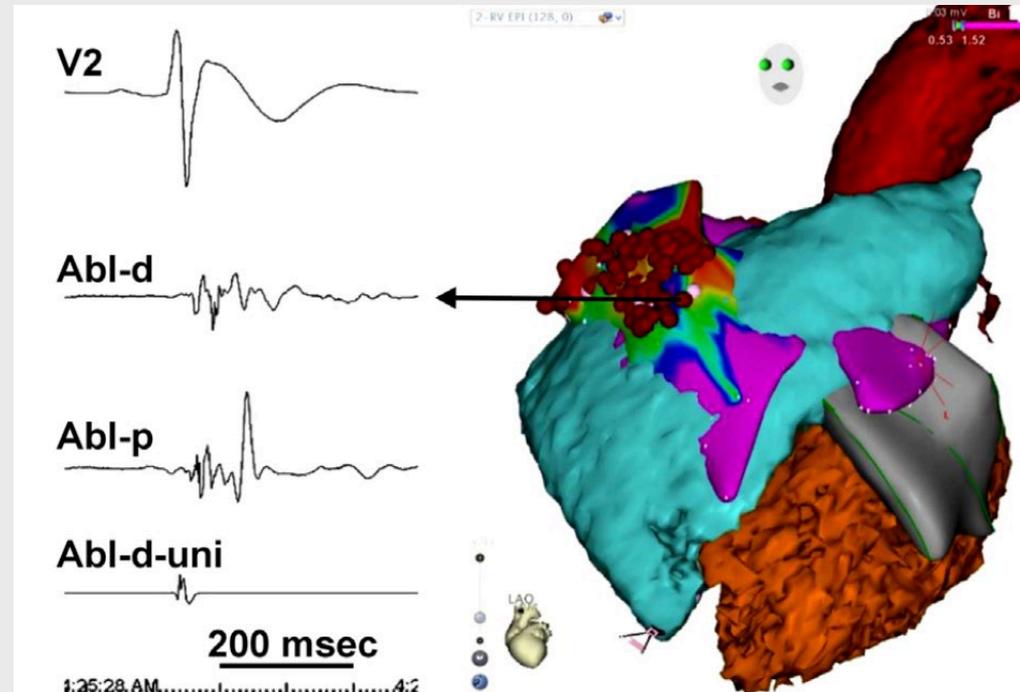
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# Brugada Syndrom – Stellenwert der Katheterablation

*Nademanee et al., Circulation. 2011;123:1270-1279*  
*Nademanee et al., Heart Rhythm 2017;14:457-461*

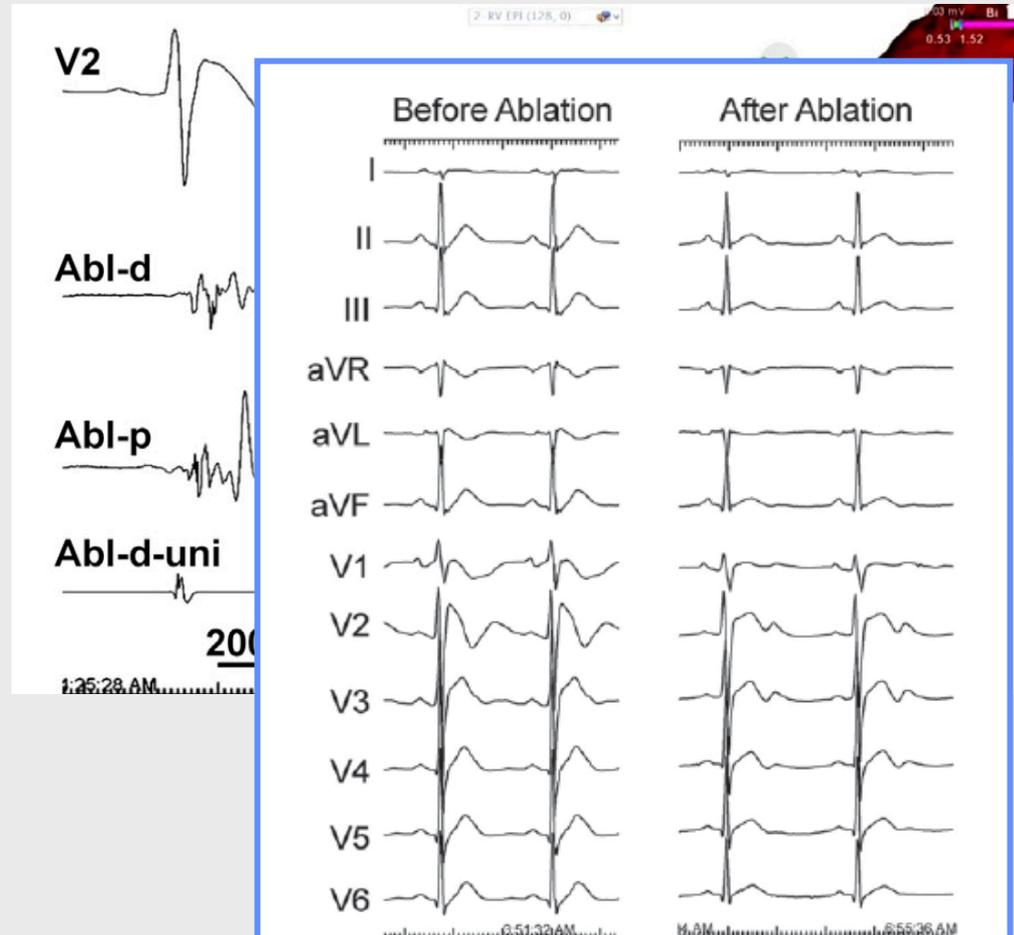
- Serie von 9 Pat. mit Typ I BrS EKG und vielen VF Episoden
- Epikardiales Mapping im RVOT zeigte abnormale, niedrig-amplitudige, fraktionierte Potentiale
- RF Ablation führte zur Nicht-Induzierbarkeit von VT/VF, verhinderte VT/VF Rezidive bei der Mehrzahl der Pat.



# Brugada Syndrom – Stellenwert der Katheterablation

*Nademanee et al., Circulation. 2011;123:1270-1279*  
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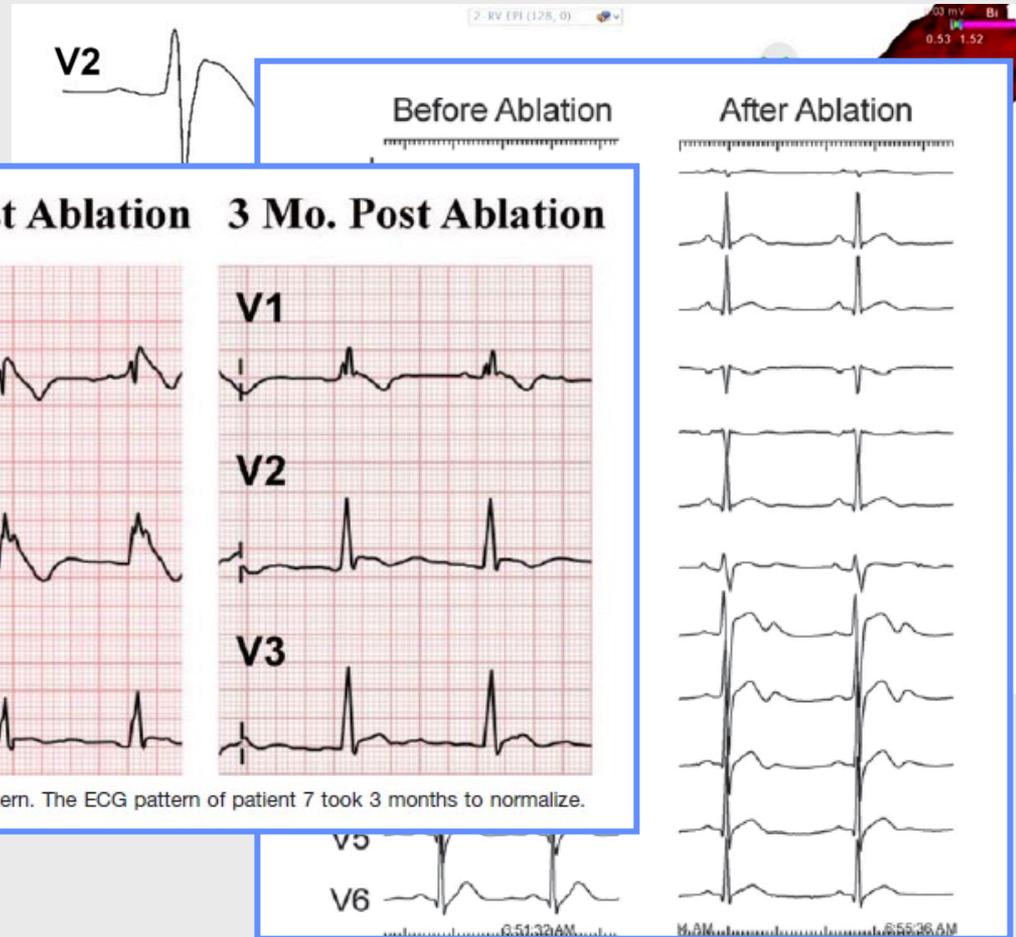
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# Brugada Syndrom – Stellenwert der Katheterablation

*Nademanee et al., Circulation. 2011;123:1270-1279*  
*Nademanee et al., Heart Rhythm 2017;14:457-461*

- Serie von 9 Pat. mit Typ I BrS EKG und vielen VF



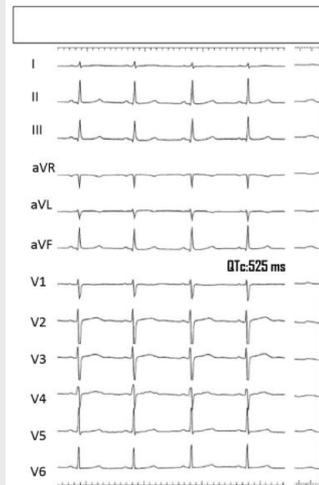
- E R ni fra
- R In ve be

# LQTS– Stellenwert der Katheterablation

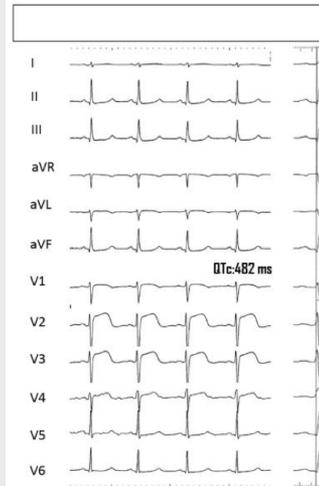
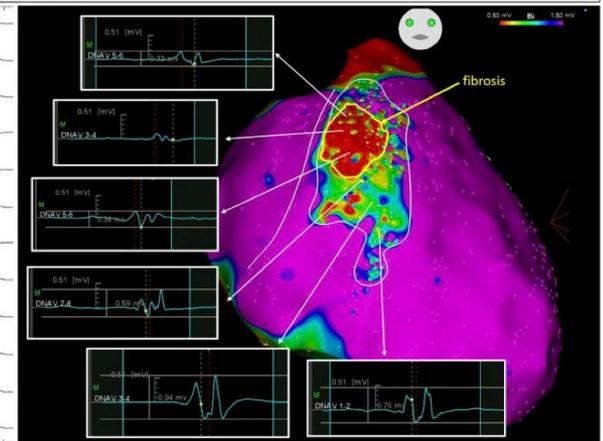
*Pappone et al., Europace 2023;5:948-955*

11 LQTS ( $44 \pm 8$  Jahre) mit im Median 4 ICD-Schocks bei VF  
 $QT_c$   $500 \pm 30$  ms  
Endo-epikardiales RV/LV Mapping mit Niedervoltage und fraktionierten Potentialen im RV epi über einer Fläche;  $16 \pm 3$  cm<sup>2</sup>.

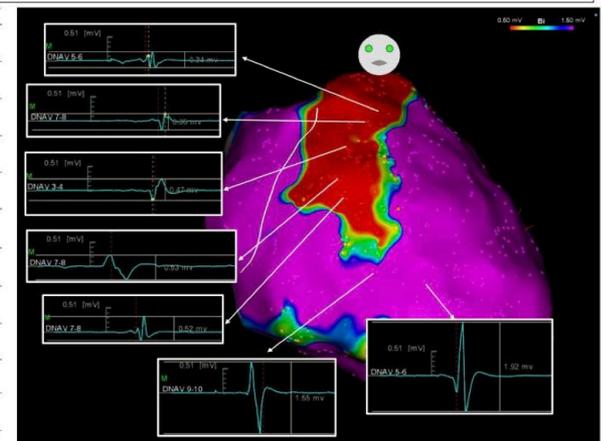
**Ablation RV epi führte zu VF Freiheit in 12 Monaten  
Follow-up und signifikanter  $QT_c$  Verkürzung ( $462 \pm 24$  ms )**



Before Ablation



After Ablation



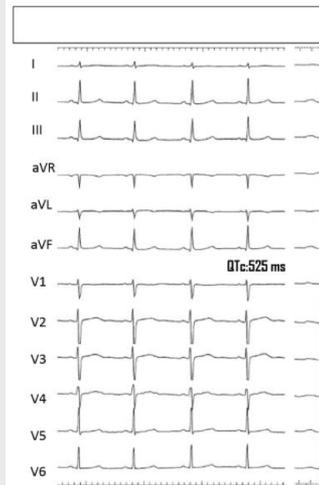
# Right ventricular epicardial arrhythmogenic substrate in long-QT syndrome patients at risk of sudden death

## Catheter ablation

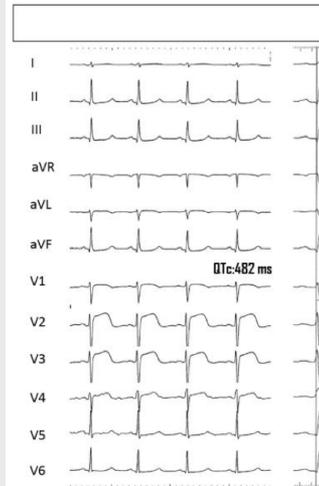
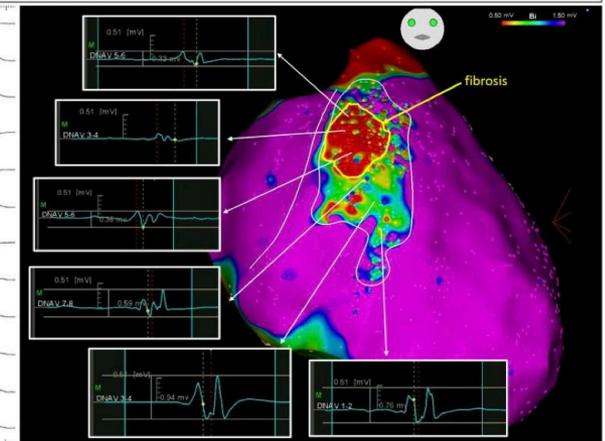
*Pappone et al., Europace 2023;5:948-955*

11 LQTS (44 ± 8 Jahre) mit im Median 4 ICD-Schocks bei VF  
QT<sub>c</sub> 500 ± 30 ms  
Endo-epikardiales RV/LV Mapping mit Niedervoltage und fraktionierten Potentialen im RV epi über einer Fläche; 16 ± 3 cm<sup>2</sup>.

**Ablation RV epi führte zu VF Freiheit in 12 Monaten  
Follow-up und signifikanter QT<sub>c</sub> Verkürzung (462 ± 24 ms)**



Before Ablation



After Ablation

