ARVC-SELBSTHILFE TRIFFT FACHWISSEN

Ärzte, Patienten und Wissenschaftler im Dialog

Samstag, 17. Juni 2023
9.30 - 17 Uhr
im St. Vinzenz Haus, LMU München

PROGRAMM

European Reference Network

*Eine Veranstaltung des **ARVC-Selbsthilfe e.V.** in Zusammenarbeit mit den deutschen Mitgliedern des Europäischen Referenznetzwerks **ERN GUARD-Heart** (Munich Consortium und Münster) und dem **wissenschaftlichen Beirat** des ARVC-Selbsthilfe e.V.*

European Reference Network
for rare or low prevalence complex diseases

- Network
  Heart Diseases
  (ERN GUARD-HEART)
- Member
  Munich consortium for rare heart disease — Deutschland

ARVC - Grundlagen

Prof. Dr. med. Stefan Kääb
LMU-Klinikum
Medizinische Klinik und Poliklinik I

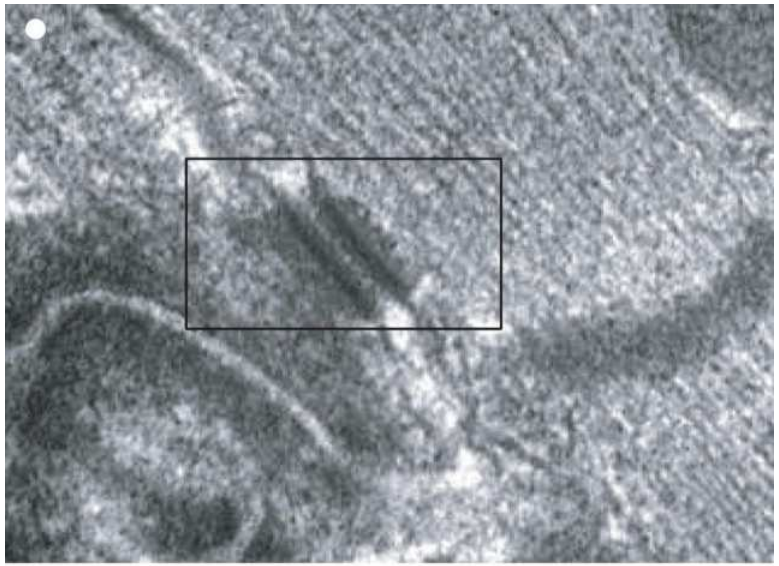
Munich Heart Alliance

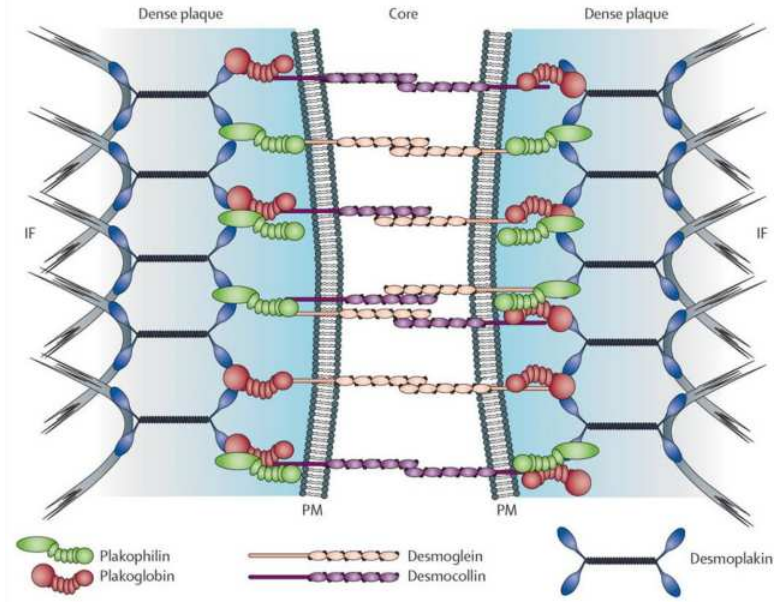
DZHK
DEUTSCHES ZENTRUM FÜR HERZ-KREISLAUF-FORSCHUNG E.V.

LMU

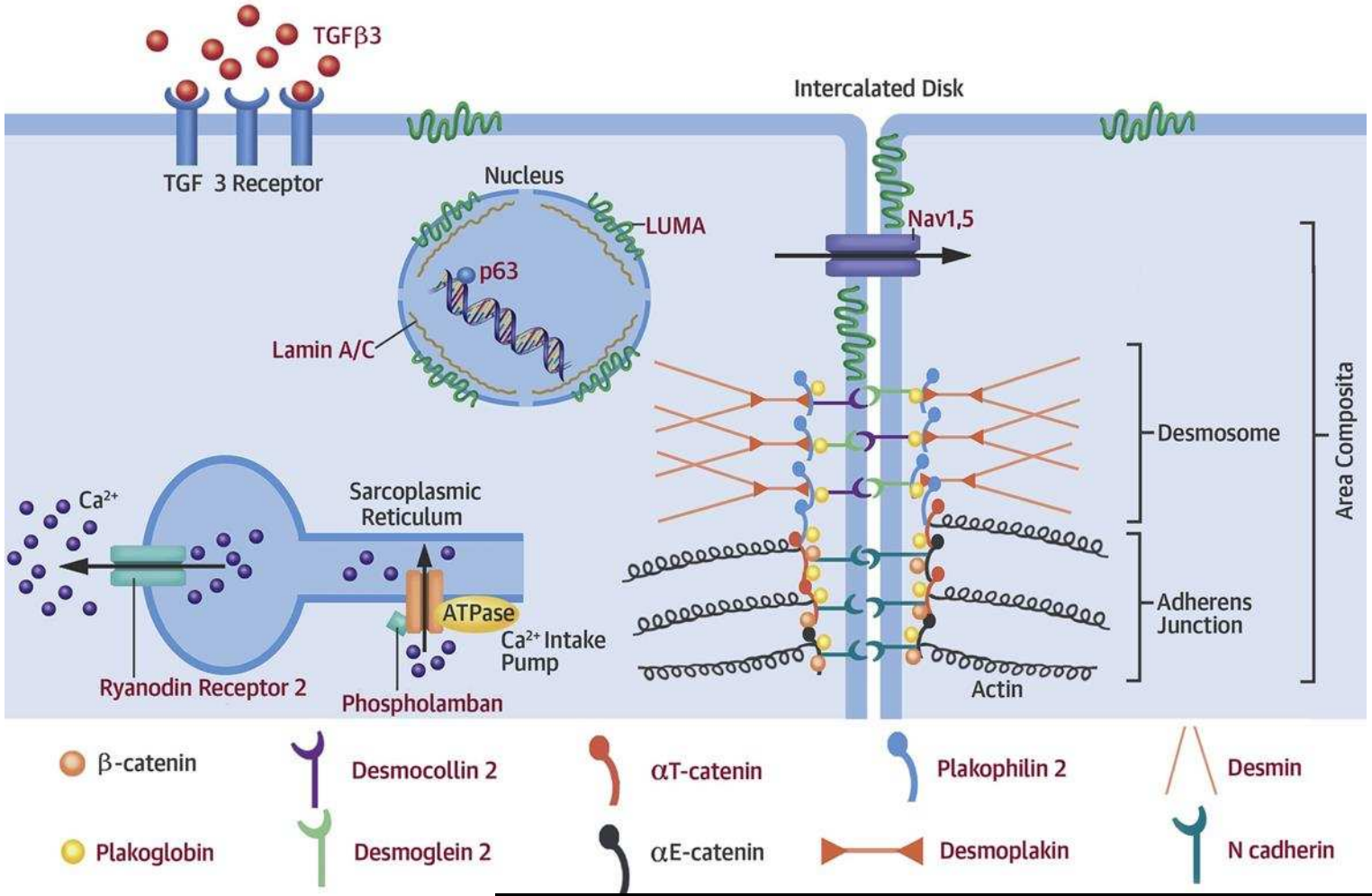
MÜNCHENER ZENTRUM FÜR SELTENE ERKRANKUNGEN (MZSE)

ARVC, a disease of the desmosome



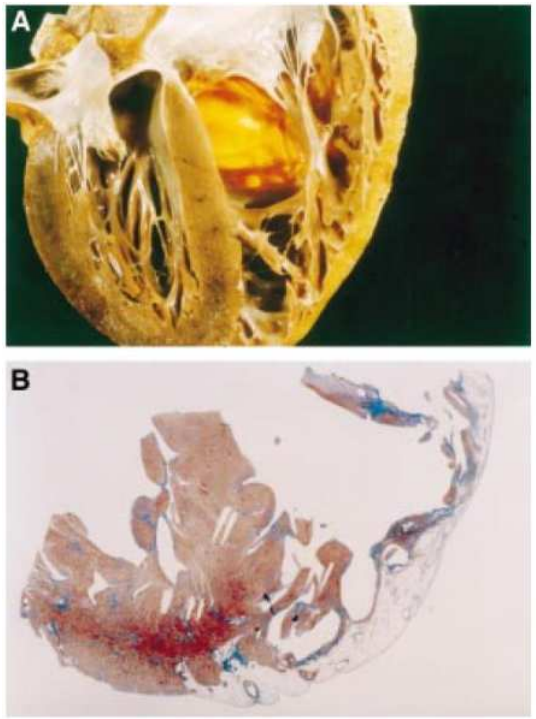


Basso C, et al. Lancet 2009

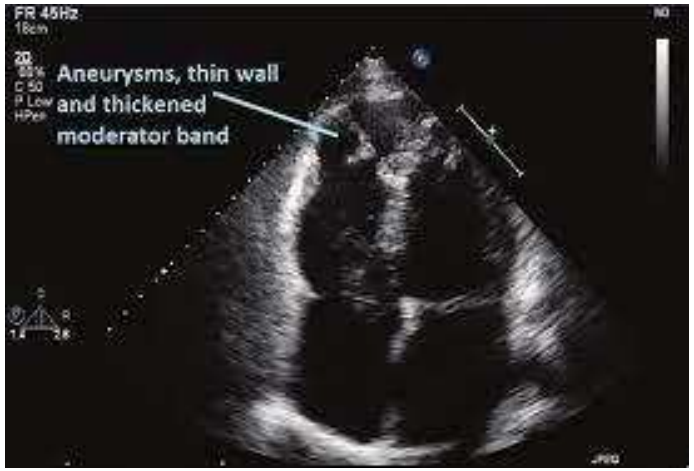


Gandjbakhch E, et al. J Am Coll Cardiol 2018

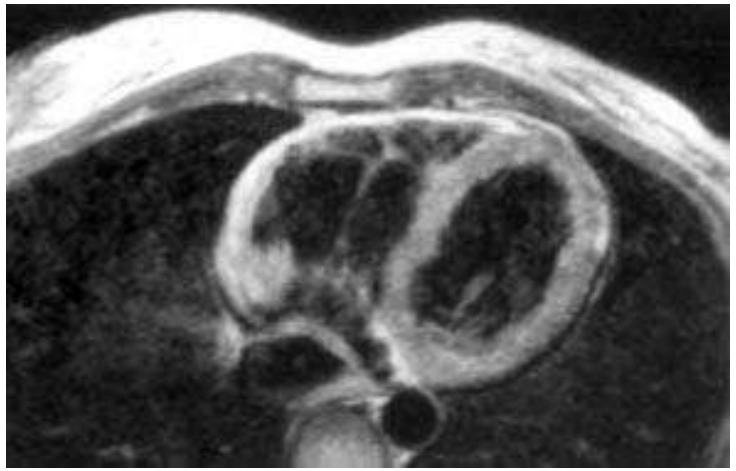
ARVC, a disease of the desmosome



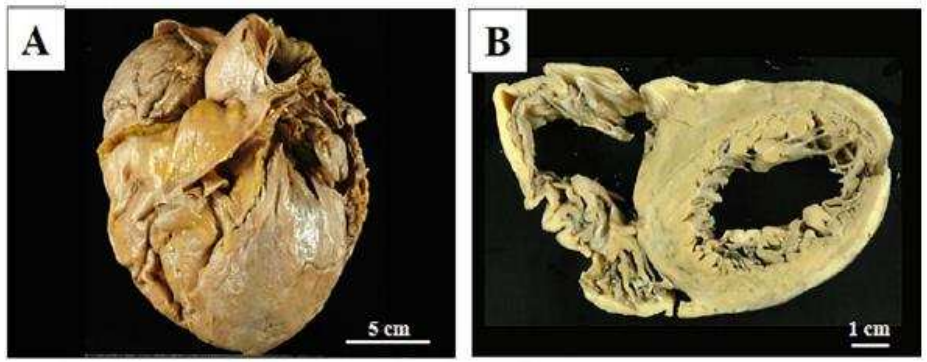
Corrado D et al., Heart 2000



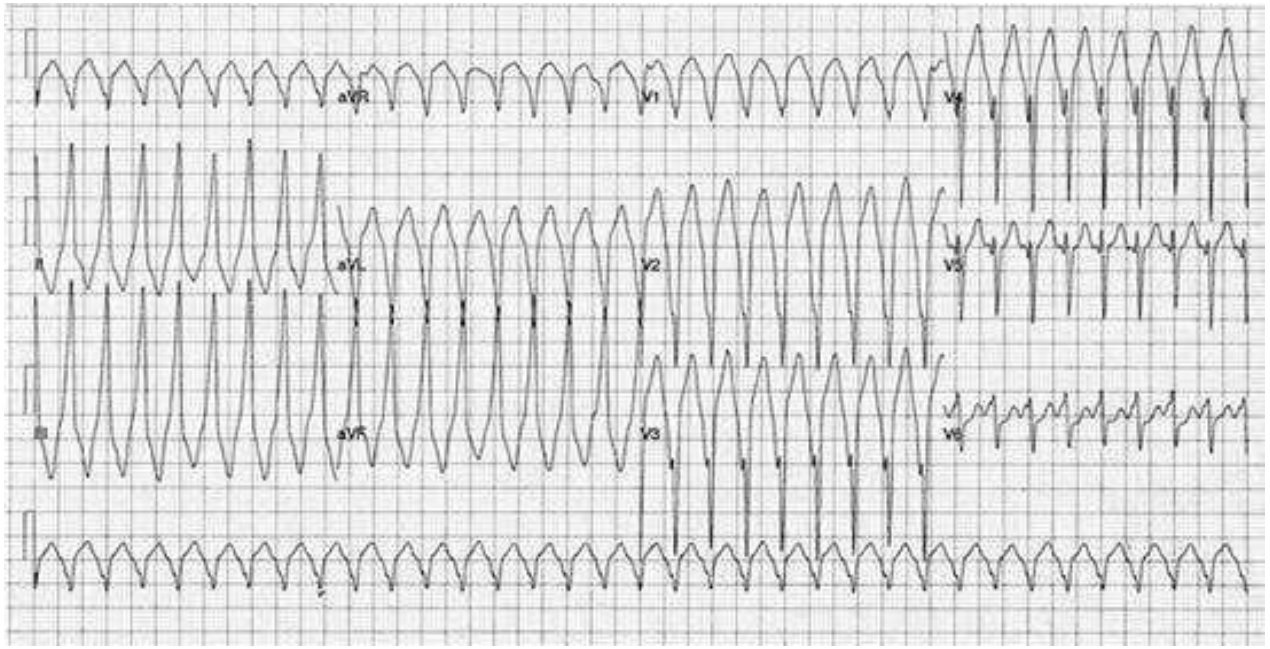
Oxborough D. et al.



Corrado D et al., Heart 2000



Yamamoto K et al., Clin. Med. Insights 2019



Diez D et al. 2008

Evolution of diagnostic criteria for ARVC/ACM

Task Force criteria (1994)

McKenna WJ et al., Br Heart J 1994

- Specificity ↑
- Sensitivity ↓
- No quantitative measurements
- Subjective criteria

Modified Task Force criteria (2010)

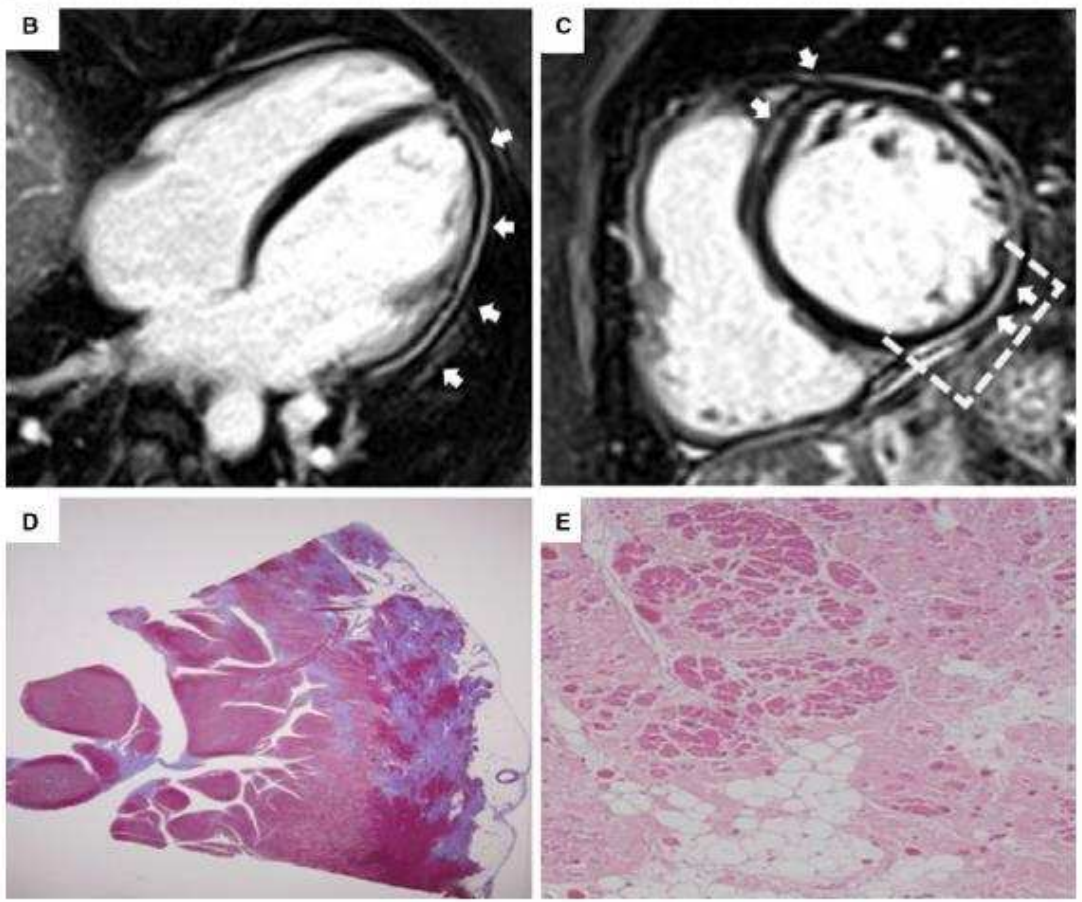
Marcus F et al., Circulation 2010

- New diagnostic methods included
- Specific definition and quantification of criteria
- Major criteria = specific for ARVC
- Additional criteria included
- Higher sensitivity, similar specificity

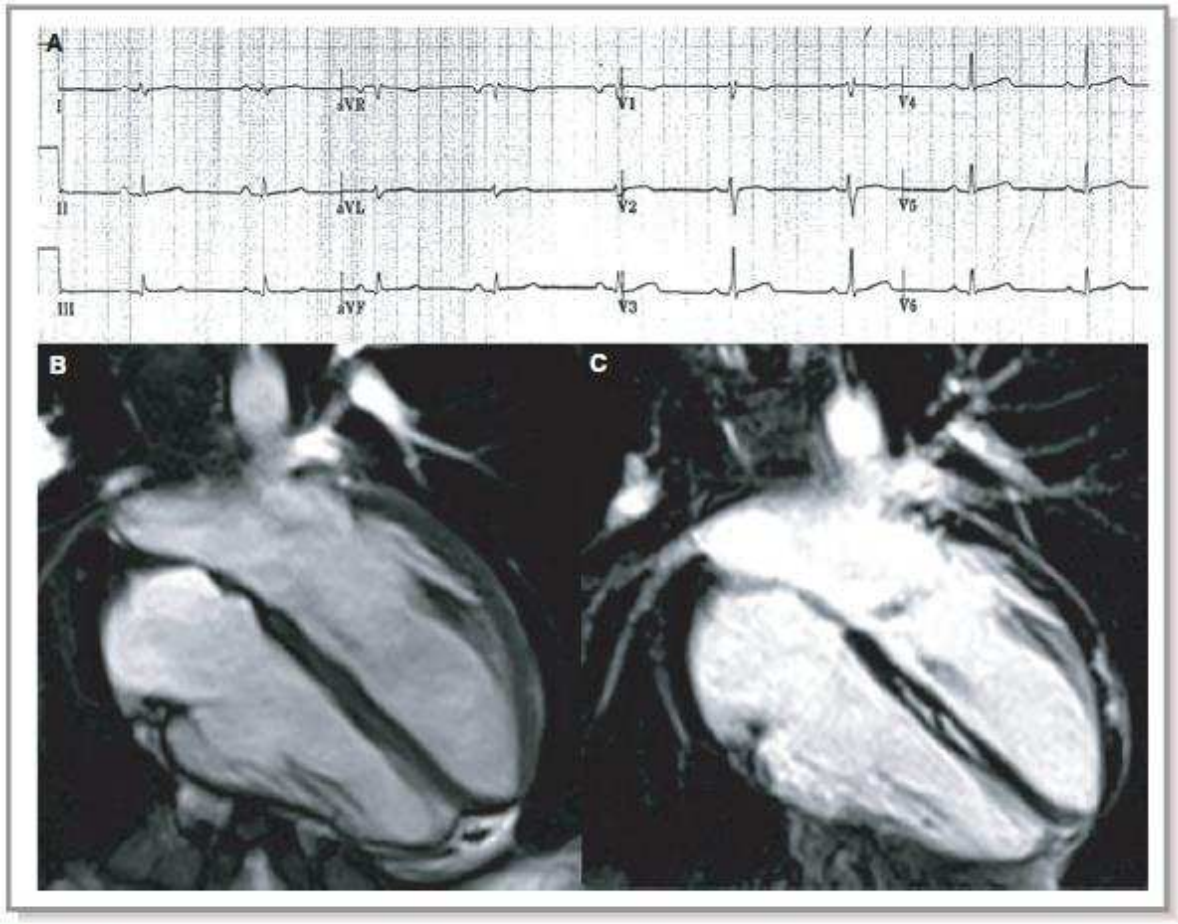
⇒ Diagnosis at early stages

⇒ Family screening

LV involvement

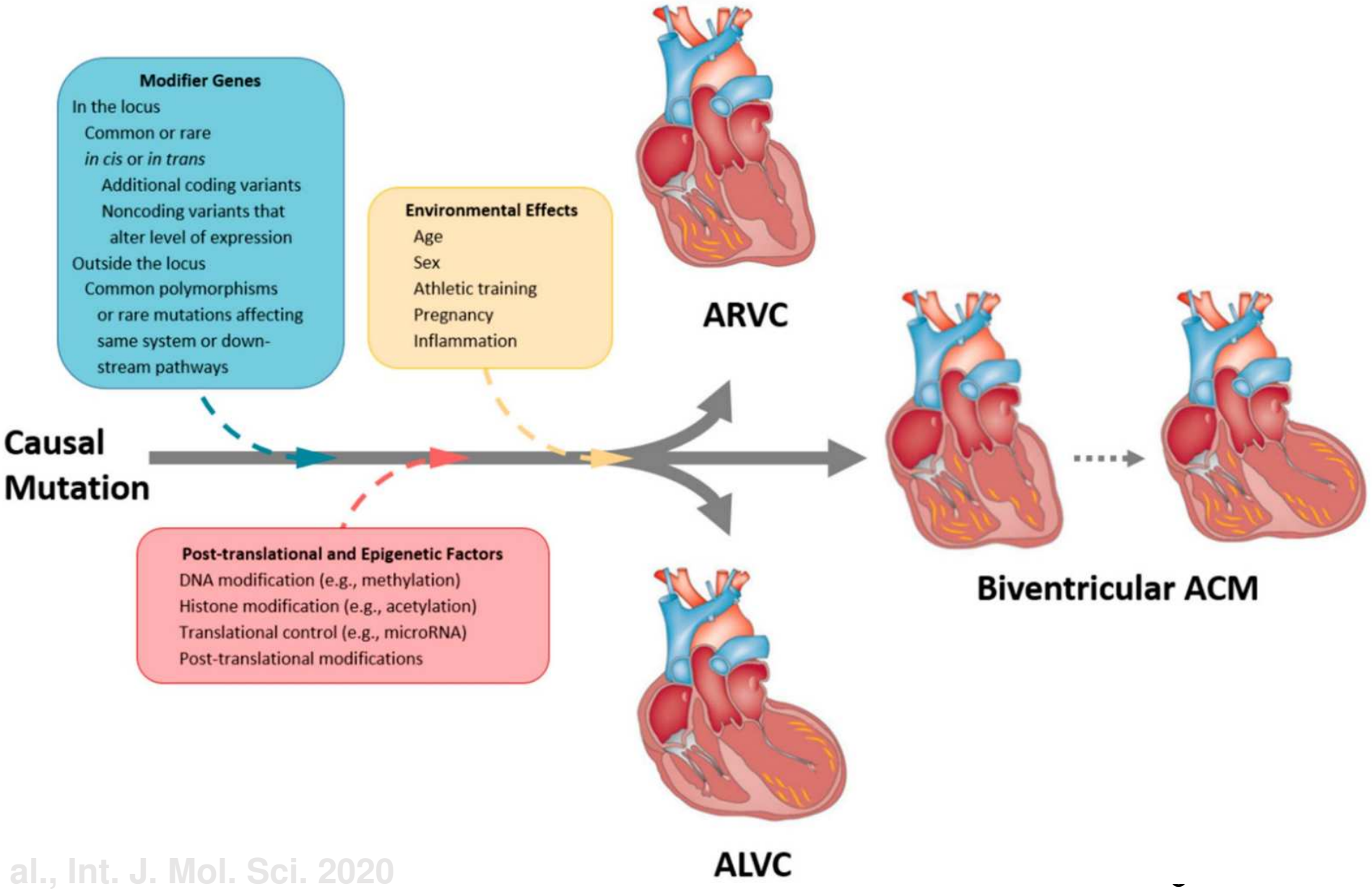


Cipriani A et al., J Am Heart Assoc. 2020



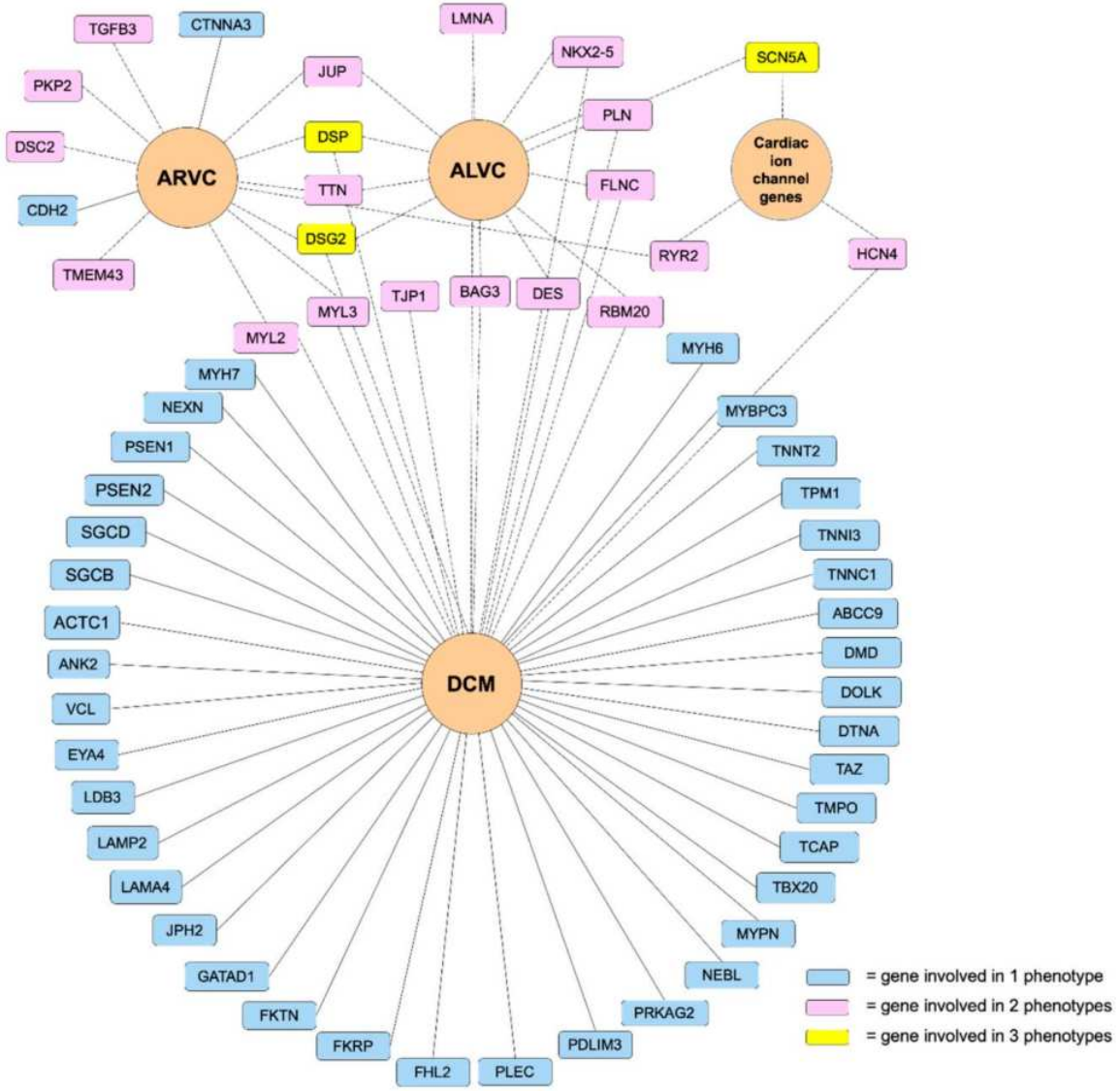
Lazzari M et al., J Am Heart Assoc. 2018

Klinische und genetische Heterogenität bei ARVC/ACM



Patel V. et al., Int. J. Mol. Sci. 2020

Klinische und genetische Heterogenität bei ARVC/ACM



Patel V. et al., Int. J. Mol. Sci. 2020

Evolution of diagnostic criteria for ARVC/ACM

LMU KLINIKUM

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⇒ Diagnosis at early stages

⇒ Family screening

Padua criteria (2020)

Corrado D et al., Int J Cardiol. 2020

- Refinement of criteria
- Inclusion of modern imaging (CE-CMRI, strain imaging)
- Inclusion of criteria for LV involvement

⇒ Evaluation of different ACM phenotypes

ARVC/ACM – Imaging Features

International Task Force Criteria 2010

ESC European Society of Cardiology
European Heart Journal (2022) **43**, 3997–4126
https://doi.org/10.1093/eurheartj/ehac262

ESC GUIDELINES

2022 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

Developed by the task force for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death of the European Society of Cardiology (ESC)

Recommendation Table 29 — Recommendations for diagnostic, risk stratification, sudden cardiac death prevention, and treatment of ventricular arrhythmias in arrhythmogenic right ventricular cardiomyopathy

Recommendations	Class[a]	Level[b]
Diagnostic evaluation and general recommendations		
In patients with suspected ARVC, CMR is recommended.[676–678]	I	B

„CMR currently provides the most accurate and reproducible measurement of atrial, biventricular global and regional systolic function, and can detect myocardial oedema, fibrosis, infiltration, and perfusion defects.“

Borgquist R, Haugaa KH, Gilljam T, Bundgaard H, Hansen J, Eschen O, et al. The diagnostic performance of imaging methods in ARVC using the 2010 task force criteria.Eur Heart J Cardiovasc Imaging 2014;15:1219–1225
European Heart Journal (2022) 43, 3997–4126

ARVC/ACM – Imaging Features

International Task Force Criteria 2010

Modified Task Force Criteria for ARVC – Diagnostic Categories Major and Minor Criteria

Definite: 2 major OR 1 major and 2 minor, OR 4 minor criteria from different categories
Borderline: 1 major and 1 minor, OR 3 minor criteria from different categories
Possible: 1 major, OR 2 minor criteria from different categories

	Major	Minor
Global or regional dysfunction and structural alterations determined by echo, MRI, or RV angiography:		
Echo	Regional RV akinesia, dyskinesia, or aneurysm and 1 of the following (end diastole): a) PLAX RVOT ≥32 mm (PLAX/BSA ≥19 mm/m^2) b) PSAX RVOT ≥36 mm (PSAX/BSA ≥21 mm/m^2) c) Fractional area change ≤33%	Regional RV akinesia, dyskinesia, or aneurysm and 1 of the following (end diastole): a) PLAX RVOT ≥29 mm to <32 mm (PLAX/BSA ≥16 to <19 mm/m^2) b) PSAX RVOT ≥32 to <36 mm (PSAX/BSA ≥18 to <21 mm/m^2) c) Fractional area change >33 to ≤40%
MRI	Regional RV akinesia or dyskinesia or dyssynchronous RV contraction and 1 of the following: a) Ratio RVEDV/BSA ≥110 mL/m^2 (male), ≥100 mL/m^2 (female) b) RVEF ≤40%	Regional RV akinesia or dyskinesia or dyssynchronous RV contraction and 1 of the following: a) Ratio RVEDV/BSA ≥100 to <110 mL/m^2 (male), ≥90 to 100 mL/m^2 (female) b) RVEF >40 to ≤45%
RV angiography	Regional RV akinesia, dyskinesia, or aneurysm	
Tissue characterization of wall		
Endomyocardial biopsy showing fibrous replacement of the RV free wall myocardium in ≥1 sample, with or without fatty replacement and with:	Residual myocytes <60% by morphometric analysis (or <50% if estimated)	Residual myocytes 60% to 75% by morphometric analysis (or 50% to 65% if estimated)
Repolarization abnormalities		
ECG	Inverted T waves in right precordial leads (V_1, V_2, and V_3) or beyond in individuals >14 years of age (in the absence of complete RBBB QRS ≥120ms)	I. Inverted T waves in leads V_1 and V_2 in individuals >14 years of age (in the absence of complete RBBB) or in V_4, V_5, or V_6. II. Inverted T waves in leads V_1, V_2, V_3, and V_4 in individuals >14 years of age in the presence of complete RBBB
Depolarization/conduction abnormalities		
ECG	Epsilon wave (reproducible low-amplitude signals between end of QRS complex to onset of the T wave) in the right precordial leads (V_1 to V_3)	I. Late potentials by SAECG in ≥1 of 3 parameters in the absence of QRS duration of ≥110ms on the standard ECG: a) Filtered QRS duration (fQRS) ≥114 ms b) Duration of terminal QRS <40 µV (low-amplitude signal duration) ≥38 ms c) Root-mean-square voltage of terminal 40 ms ≤20 µV II. Terminal activation duration of QRS ≥55 ms measured from the nadir of the S wave to the end of the QRS, including R' in V_1, V_2, or V_3 in the absence of complete RBBB
Arrhythmias		
	Nonsustained or sustained VT of LBBB with superior axis (negative or indeterminate QRS in leads II, III, and aVF and positive in lead aVL)	I. Nonsustained or sustained VT or RV outflow configuration, LBBB morphology with inferior axis (positive QRS in II, III and aVF and negative in lead aVL) or of unknown axis II. >500 ventricular extrasystoles per 24 hours (Holter)
Family history		
	I. ARVC confirmed in a first-degree relative who meets current Task Force Criteria II. ARVC confirmed pathologically at autopsy or surgery in a first-degree relative III. Identification of a pathogenetic mutation categorized as associated or probably associated with ARVC in the patient under evaluation	I. History of ARVC in a first-degree relative in whom it is not possible or practical to determine whether the family member meets current Task Force Criteria II. Premature sudden death (<35 years of age) due to suspected ARVC in a first-degree relative III. ARVC confirmed pathologically or by current Task Force Criteria in second-degree relative

Marcus et al. European Heart Journal (2010) 31, 806–814

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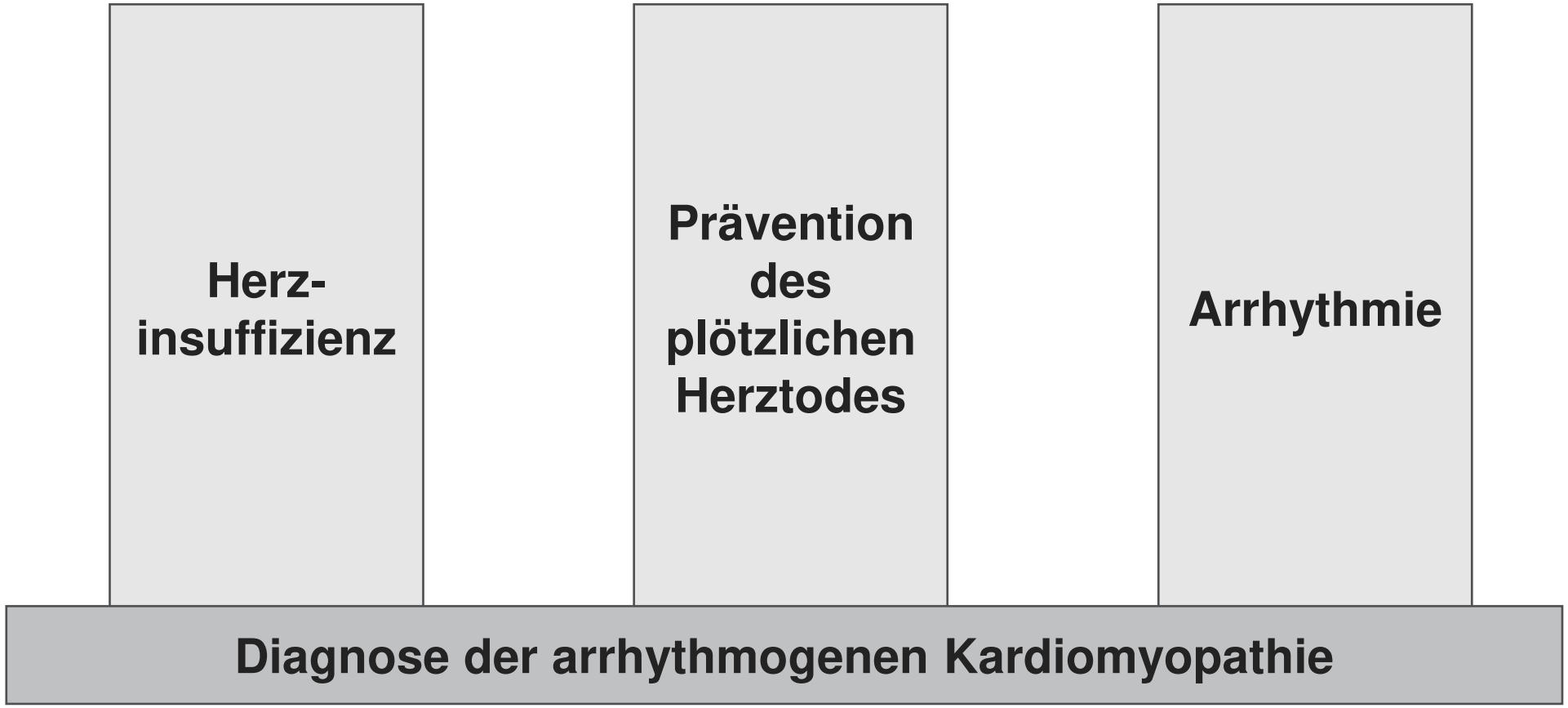
Drei Säulen der Therapie

Individuell abgestimmt nach Krankheitsverlauf und Symptomatik



PD Dr. Moritz Sinner, LMU München

Therapie der Arrhythmie: Katheterablation

IIa	B-NR
IIa	B-NR
IIa	C-EO

Bei Patienten mit ACM und wiederholter, anhaltender, monomorpher VT,
trotz Amiodaron oder die Amiodaron nicht tolerieren
Soll eine Katheterablation erwogen werden um VTs und ICD Schocks zu reduzieren

Bei Patienten mit ACM und wiederholter, symptomatischer, anhaltender VT,
trotz antiarrhythmischer Medikation oder bei Unverträglichkeit antiarrhythmischer Medikation
Soll eine kombiniert endo- / epikardiale Katheterablation erwogen werden

Bei symptomatischen Patienten mit ACM und hoher VES Last oder nsVT,
Bei welchen Betablocker oder antiarrhythmisch Medikation
Nicht vertragen werden oder unwirksam sind
Soll eine kombiniert endo- / epikardiale Katheterablation erwogen werden

Towbin JA et al. 2019 HRS ACM expert consensus statement. Heart Rhythm 2019;16:e301-e372

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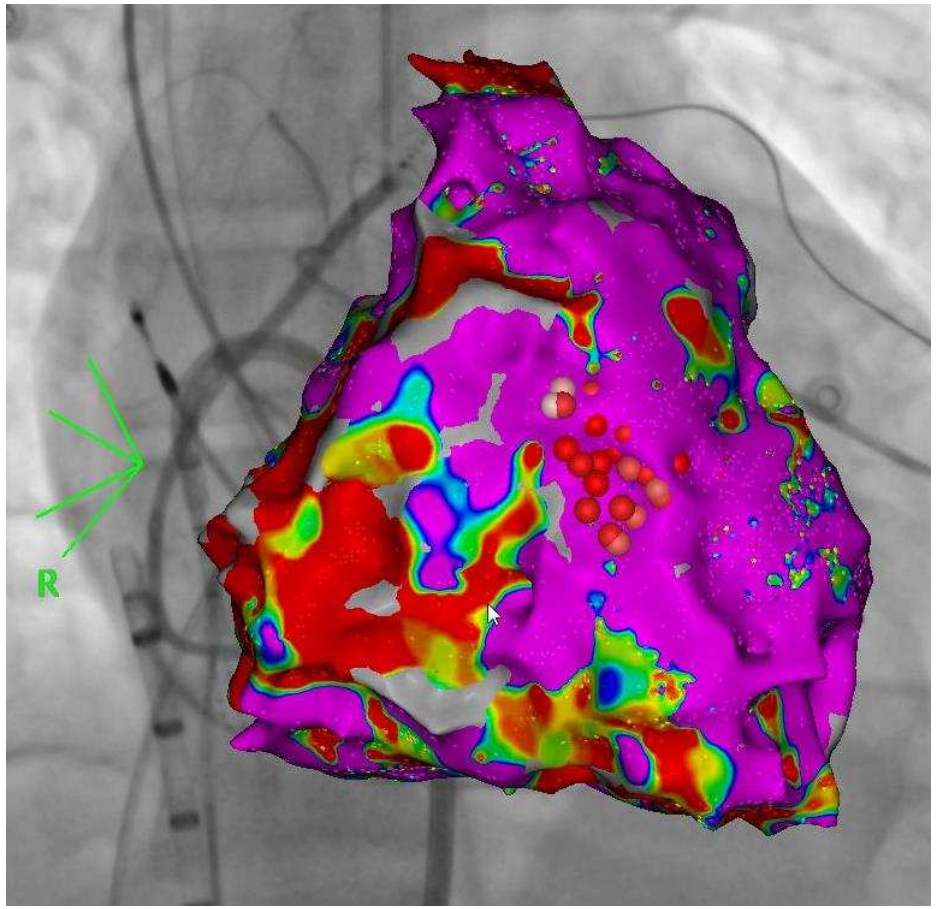
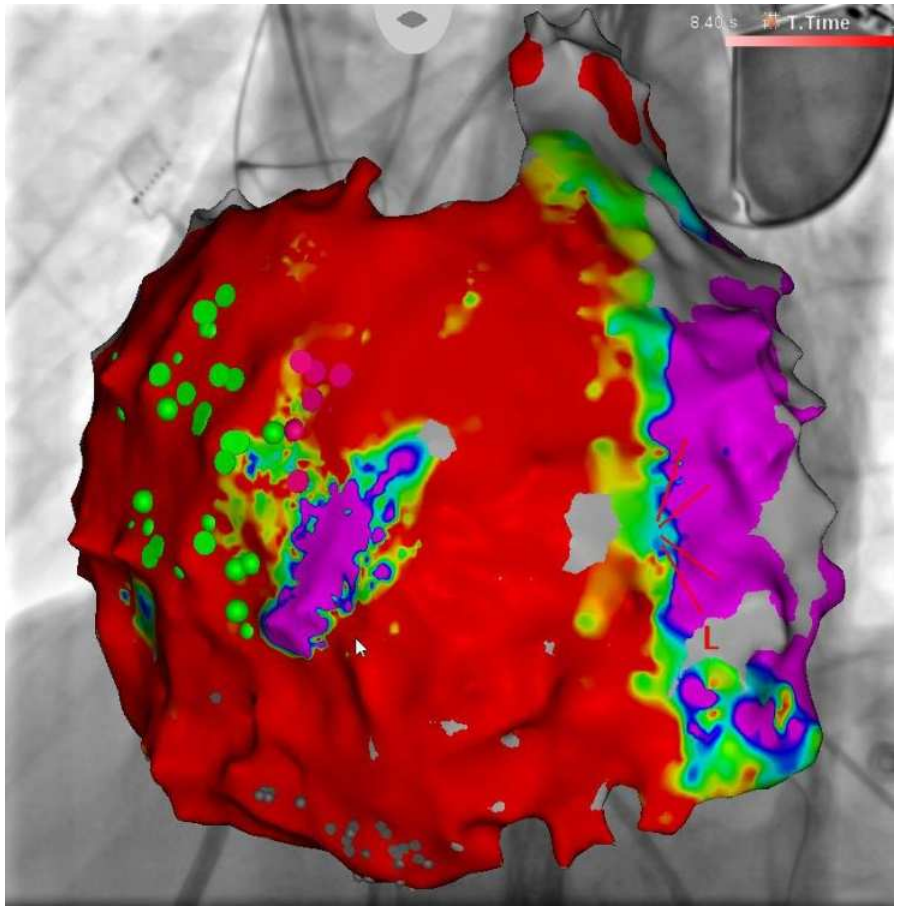
Therapie der Arrhythmie: Fallbeispiel



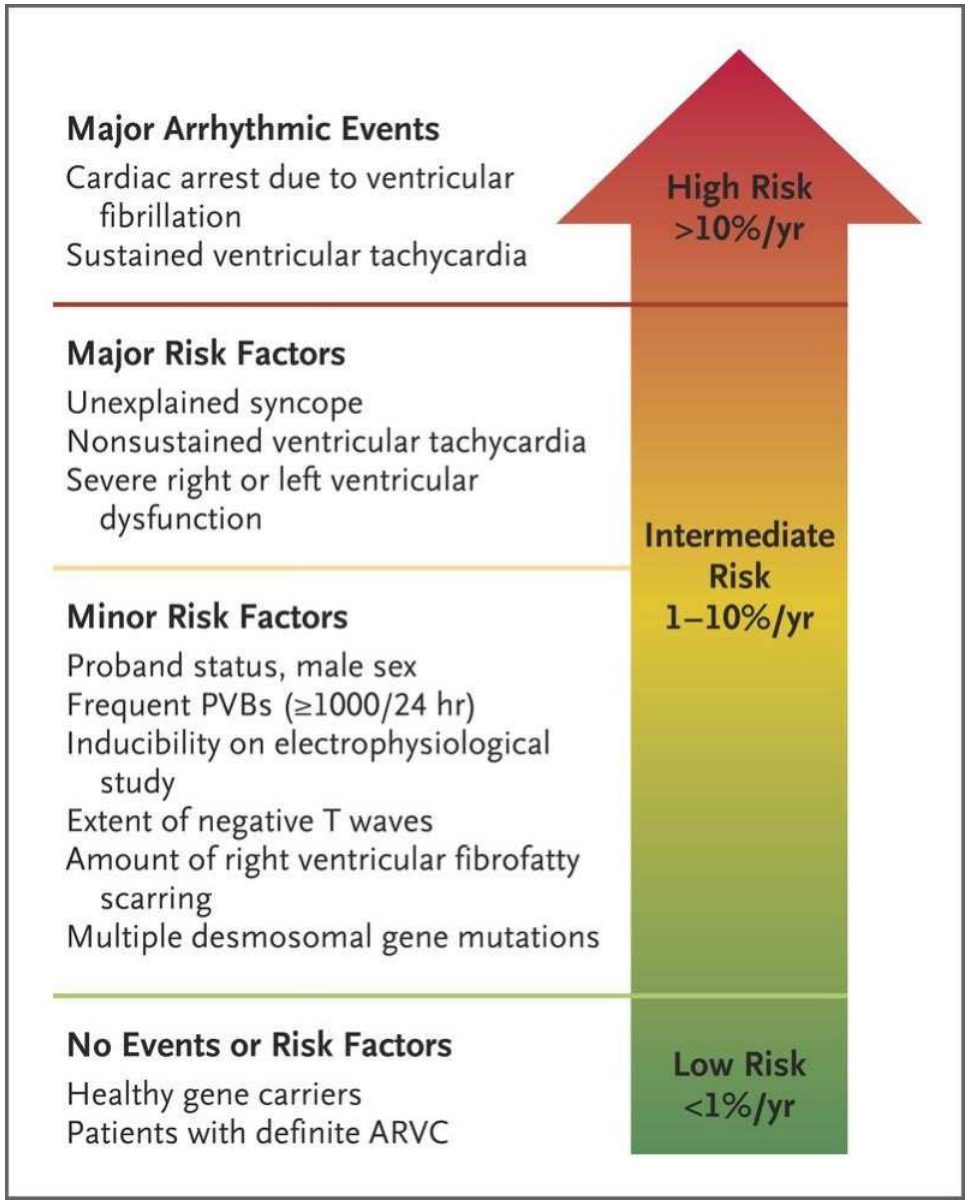
endokardial



epikardial

PD Dr. Moritz Sinner, LMU München

Risk Prediction in ARVC



Corrado D, et al NEJM 2017

Herzlichen Dank!

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