Diagnose von ARVC mit Schwerpunkt MRT

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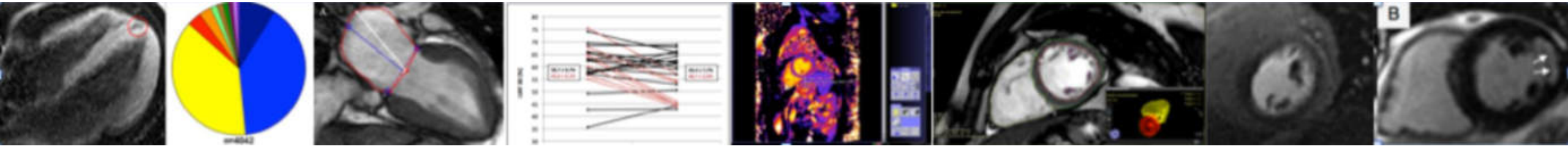


CHARITÉ UNIVERSITÄTSMEDIZIN BERLIN
MDC MAX-DELBRÜCK-CENTRUM FÜR MOLEKULARE MEDIZIN IN DER HELMHOLTZ-GEMEINSCHAFT
BIH Berlin Institute of Health Charité & MDC
DZHK DEUTSCHES ZENTRUM FÜR HERZ-KREISLAUF-FORSCHUNG E.V.
Helios

Dilemma und Chance

Circulation

JOURNAL OF THE AMERICAN HEART ASSOCIATION

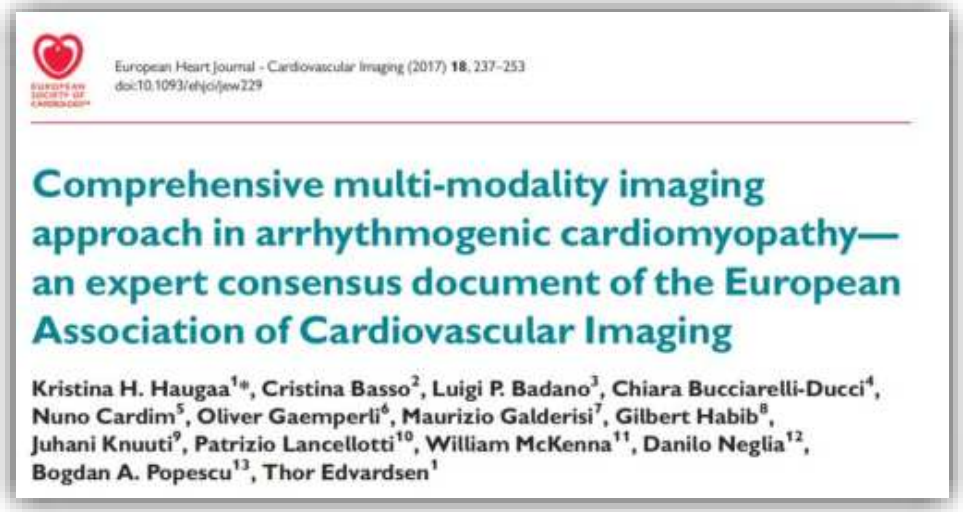
American Heart Association. Learn and Live.

Diagnosis of Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia. Proposed Modification of the Task Force Criteria

Frank I. Marcus, William J. McKenna, Duane Sherrill, Cristina Basso, Barbara Bauce, David A. Bluemke, Hugh Calkins, Domenico Corrado, Moniek G.P.J. Cox, James P. Daubert, Guy Fontaine, Kathleen Gear, Richard Hauer, Andrea Nava, Michael H. Picard, Nikos Protonotarios, Jeffrey E. Saffitz, Danita M. Yoerger Sanborn, Jonathan S. Steinberg, Harikrishna Tandri, Gaetano Thiene, Jeffrey A. Towbin, Adalena Tsatsopoulou, Thomas Wichter and Wojciech Zareba

Circulation published online Feb 19, 2010;

DOI: 10.1161/CIRCULATIONAHA.108.840827

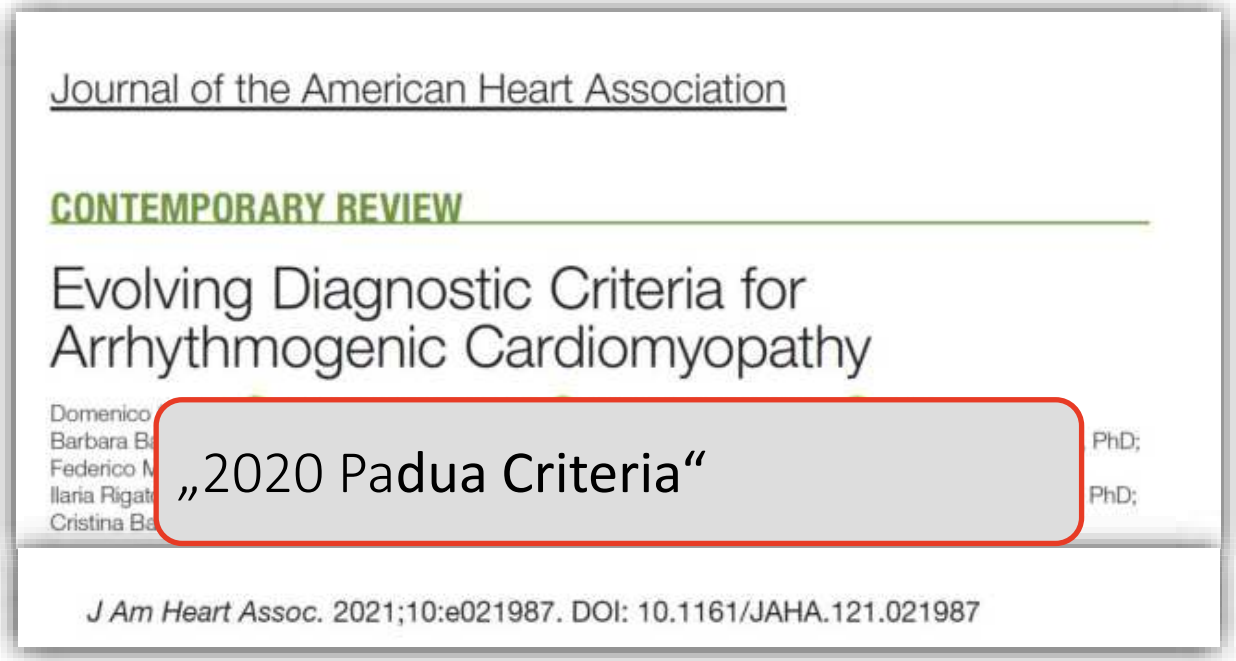


European Heart Journal - Cardiovascular Imaging (2017) **18**, 237–253
doi:10.1093/ehjci/jew229

Comprehensive multi-modality imaging approach in arrhythmogenic cardiomyopathy—an expert consensus document of the European Association of Cardiovascular Imaging

Kristina H. Haugaa[1]*, Cristina Basso[2], Luigi P. Badano[3], Chiara Bucciarelli-Ducci[4], Nuno Cardim[5], Oliver Gaemperli[6], Maurizio Galderisi[7], Gilbert Habib[8], Juhani Knuuti[9], Patrizio Lancellotti[10], William McKenna[11], Danilo Neglia[12], Bogdan A. Popescu[13], Thor Edvardsen[1]

Diagnose - mehrere Kriterien Score



Journal of the American Heart Association

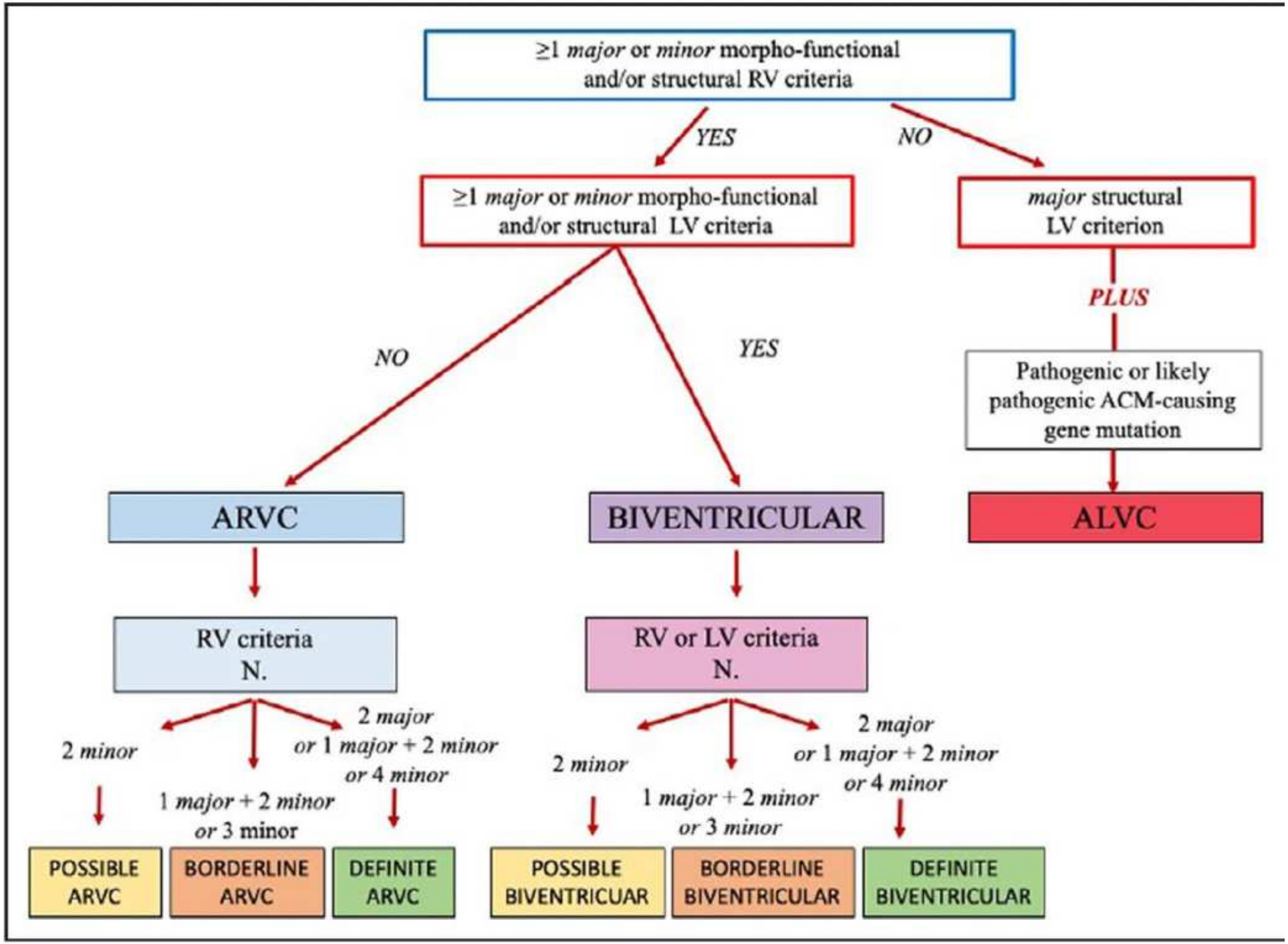
CONTEMPORARY REVIEW

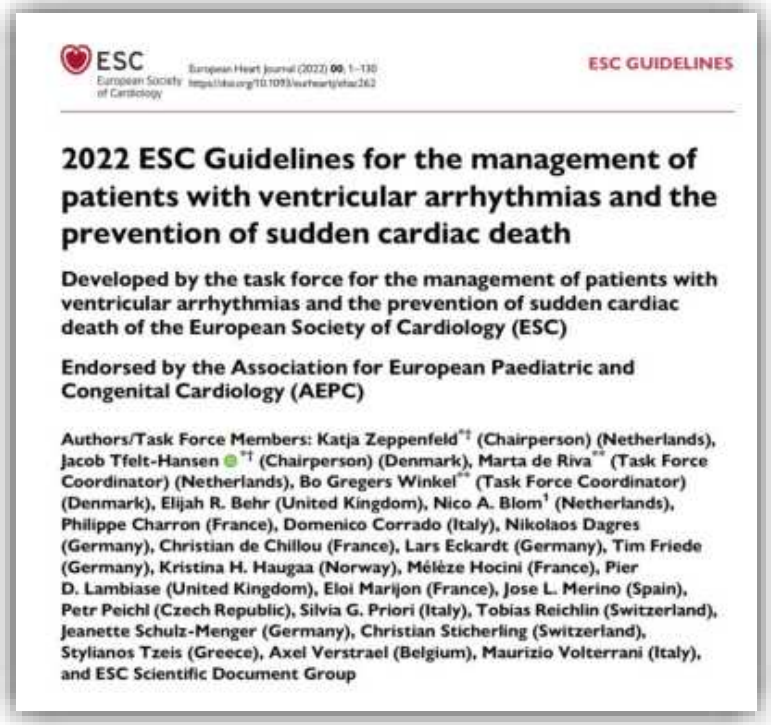
Evolving Diagnostic Criteria for Arrhythmogenic Cardiomyopathy

Domenico
Barbara Ba
Federico M
Ilaria Rigat
Cristina Ba
PhD;
PhD;

„2020 Padua Criteria“

J Am Heart Assoc. 2021;10:e021987. DOI: 10.1161/JAHA.121.021987


≥1 major or minor morpho-functional and/or structural RV criteria
YES
NO
≥1 major or minor morpho-functional and/or structural LV criteria
major structural LV criterion
PLUS
Pathogenic or likely pathogenic ACM-causing gene mutation
NO
YES
ARVC
BIVENTRICULAR
ALVC
RV criteria N.
RV or LV criteria N.
2 minor
1 major + 2 minor or 3 minor
2 major or 1 major + 2 minor or 4 minor
2 minor
1 major + 2 minor or 3 minor
2 major or 1 major + 2 minor or 4 minor
POSSIBLE ARVC
BORDERLINE ARVC
DEFINITE ARVC
POSSIBLE BIVENTRICUAR
BORDERLINE BIVENTRICULAR
DEFINITE BIVENTRICULAR



ESC European Society of Cardiology

European Heart Journal (2022) 00, 1–130 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)

Endorsed by the Association for European Paediatric and Congenital Cardiology (AEPC)

Authors/Task Force Members: Katja Zeppenfeld*† (Chairperson) (Netherlands), Jacob Tfelt-Hansen*† (Chairperson) (Denmark), Marta de Riva** (Task Force Coordinator) (Netherlands), Bo Gregers Winkel** (Task Force Coordinator) (Denmark), Elijah R. Behr (United Kingdom), Nico A. Blom† (Netherlands), Philippe Charron (France), Domenico Corrado (Italy), Nikolaos Dagres (Germany), Christian de Chillou (France), Lars Eckardt (Germany), Tim Friede (Germany), Kristina H. Haugaa (Norway), Méléze Hocini (France), Pier D. Lambiase (United Kingdom), Eloi Marijon (France), Jose L. Merino (Spain), Petr Peichl (Czech Republic), Silvia G. Priori (Italy), Tobias Reichlin (Switzerland), Jeanette Schulz-Menger (Germany), Christian Sticherling (Switzerland), Stylianos Tzeis (Greece), Axel Verstrael (Belgium), Maurizio Volterrani (Italy), and ESC Scientific Document Group

ARVC

In patients with suspected ARVC, CMR is recommended.	I
In patients with a suspected or definite diagnosis of ARVC, genetic counselling and testing are recommended.	I
ICD implantation should be considered in symptomatic[d] patients with definite ARVC, moderate right or left ventricular dysfunction, and either NSVT or inducibility of SMVT at PES.	IIa
In ARVC patients with indication for ICDs, a device with the capability of ATP programming for SMVT up to high rates	IIa

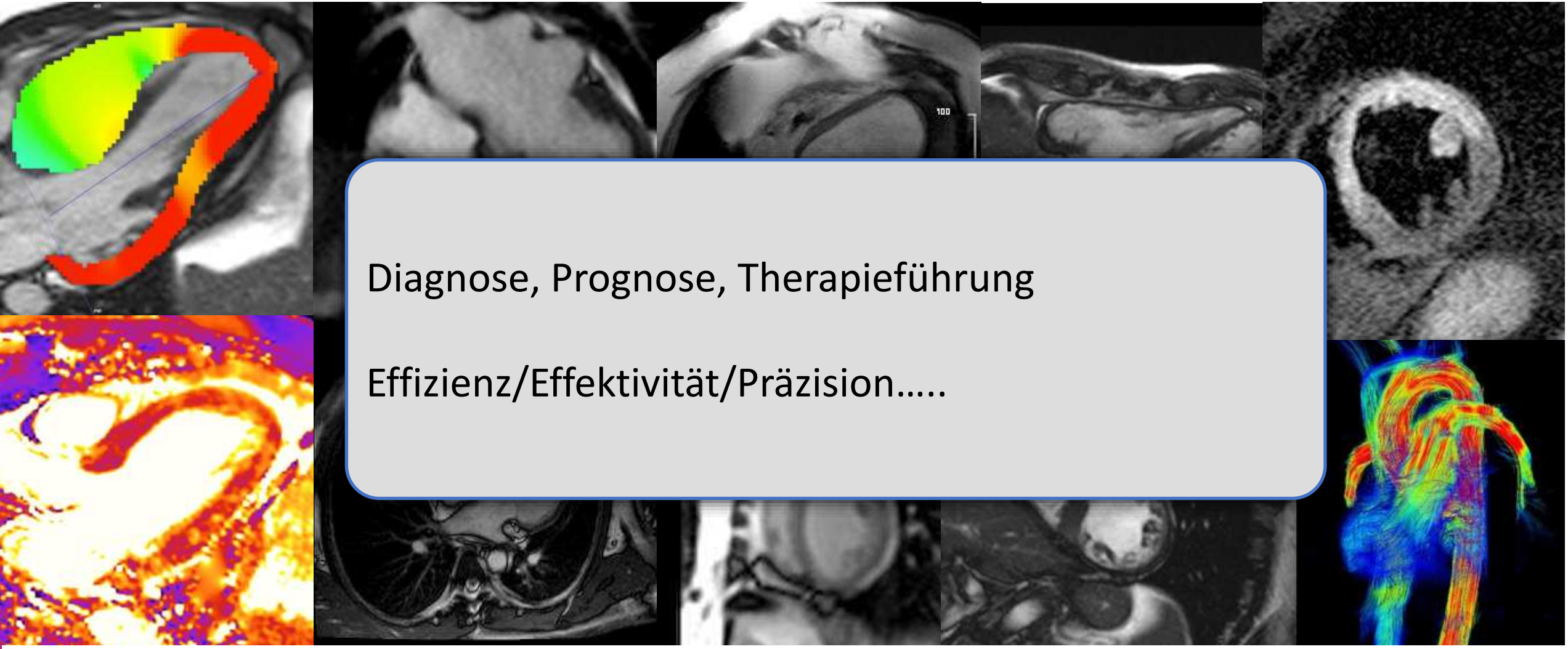
Recommendations for diagnostic, risk stratification, sudden cardiac death prevention and treatment of ventricular arrhythmia in arrhythmogenic right ventricular cardiomyopathy

In patients with suspected ARVC, CMR is recommended.	I	B

phenotype.

Beta-blocker therapy may be considered in all patients with a definite diagnosis of ARVC.	IIb
In patients with ARVC and symptoms highly suspicious for VA, PES may be considered for risk stratification.	IIb

Auswahl der Kardio-MRT Techniken



Diagnose, Prognose, Therapieführung

Effizienz/Effektivität/Präzision…..

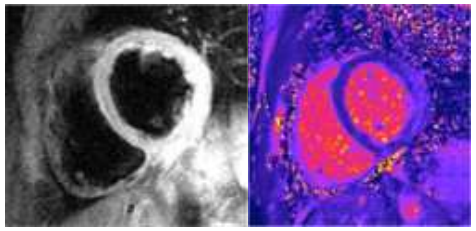
Kardio-MRT - gezieltes Protokoll nach Fragestellung notwendig

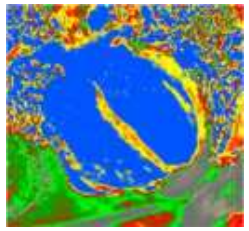
Erkrankungsaktivität

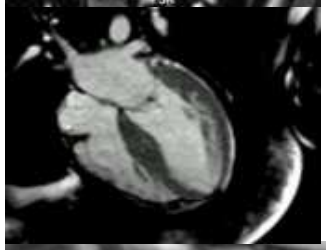
native

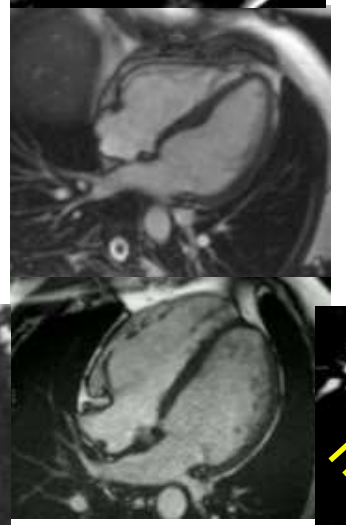
T2w/T2map

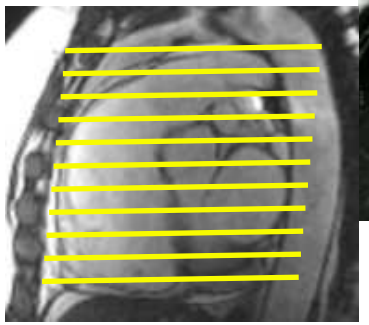
T1map

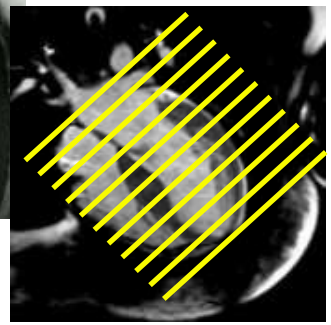










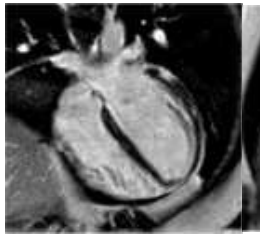


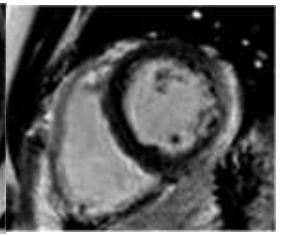
Prognose

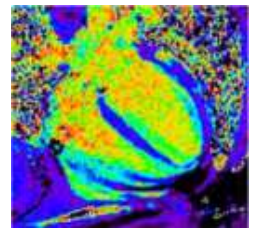
post contrast media

LGE

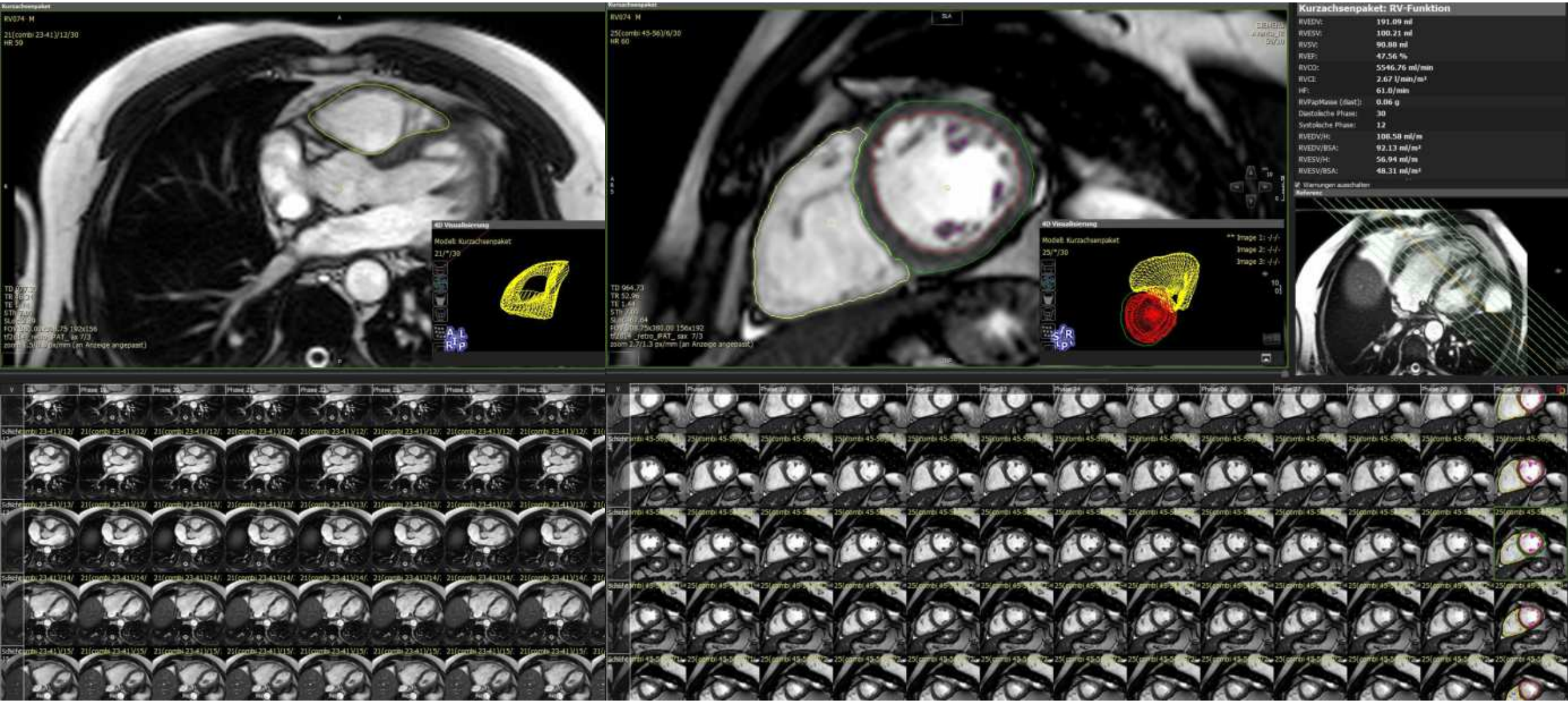
ECV







Präzise Quantifizierung beider Ventrikel



Schulz-Menger et al JCMR 2020 SCMR Guidelines

A(R)VC Funktion und Wandbewegungsstörungen

„2020 Padua Criteria“

Table 1. Comparison of 2010 TF Criteria and 2020 International Criteria for

Categ	
I. Glob or regi dysfur and st alterat	- Fractional area change ≤33% *By MRI:* • Regional RV akinesia or dyskinesia or dyssynchronous RV contraction *and* one of the following: - Ratio of RV end-diastolic volume to BSA: ≥110 mL/m² (male) or ≥100 ml (female) - *or* RV ejection fraction ≤40% *By RV angiography:* • Regional RV akinesia, dyskinesia, or aneurysm *Minor* *By 2D echocardiogram:* • Regional RV akinesia or dyskinesia *and* one of the following (end diastol - PLAX RVOT ≥29–<32 mm; (corrected for body size [PLAX/BSA] ≥16–<19 mm/m²) - PSAX RVOT ≥32–<36 mm; (corrected for body size [PSAX/BSA] ≥18–<21 mm/m²) - or fractional area change >33%–≤40% *By MRI:* • Regional RV akinesia or dyskinesia or dyssynchronous RV contraction a one of the following: - Ratio of RV end-diastolic volume to BSA ≥100 to <110 mL/m² (male) or ≥90 to <100 mL/m² (female) - *or* RV ejection fraction >40% to ≤45%

2020 International criteria

…graphy:
…r bulging

- Global RV dilatation (increase of RV EDV according to the imaging test specific nomograms for age, sex, and BSA)

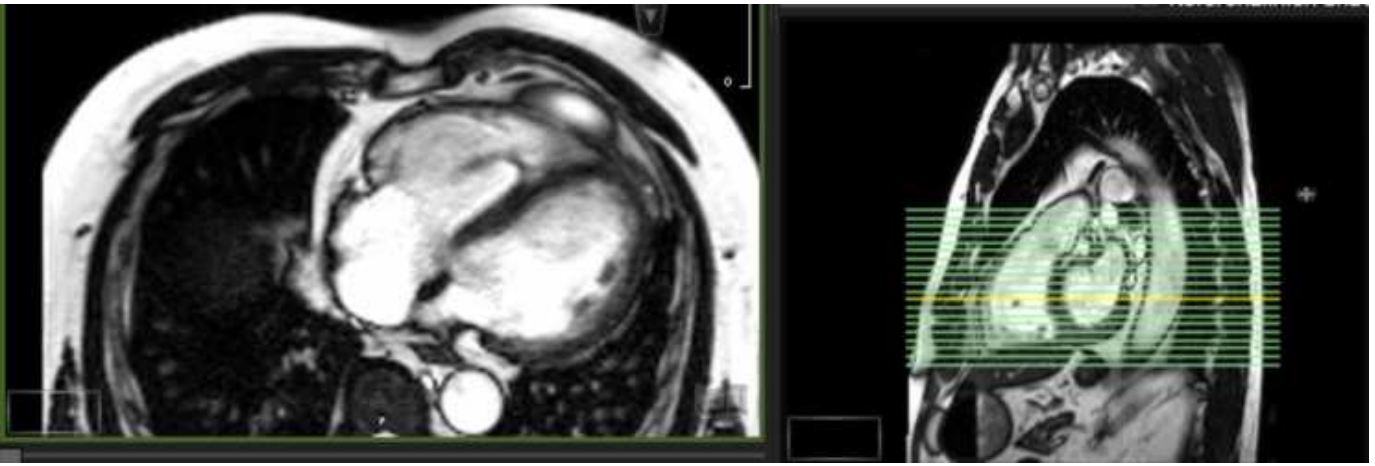
or

- Global RV systolic dysfunction (reduction of RV EF according to the imaging test specific nomograms for age and sex)

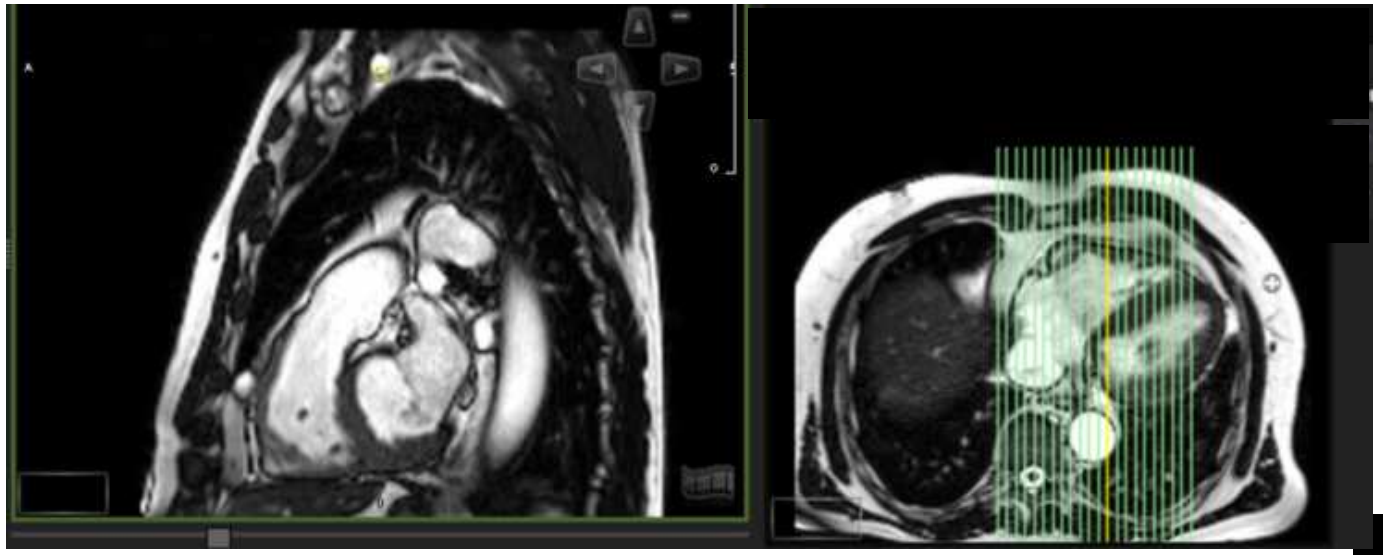
Minor

By 2D echocardiogram, CMR, or angiography:

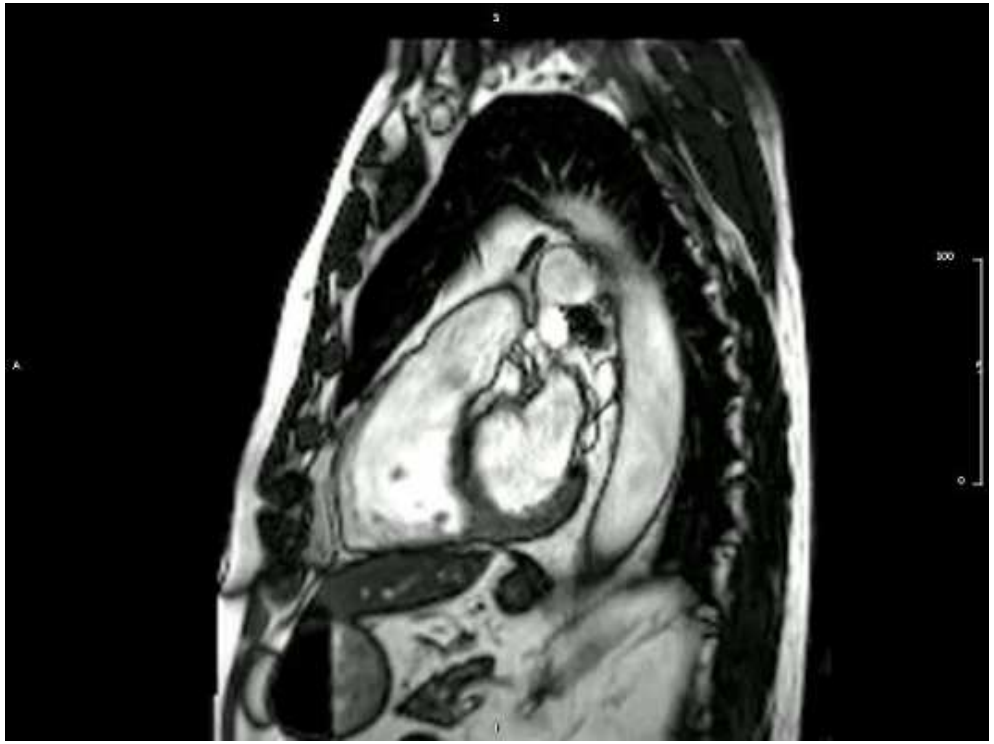
- Regional RV akinesia, dyskinesia or aneurysm of RV free wall

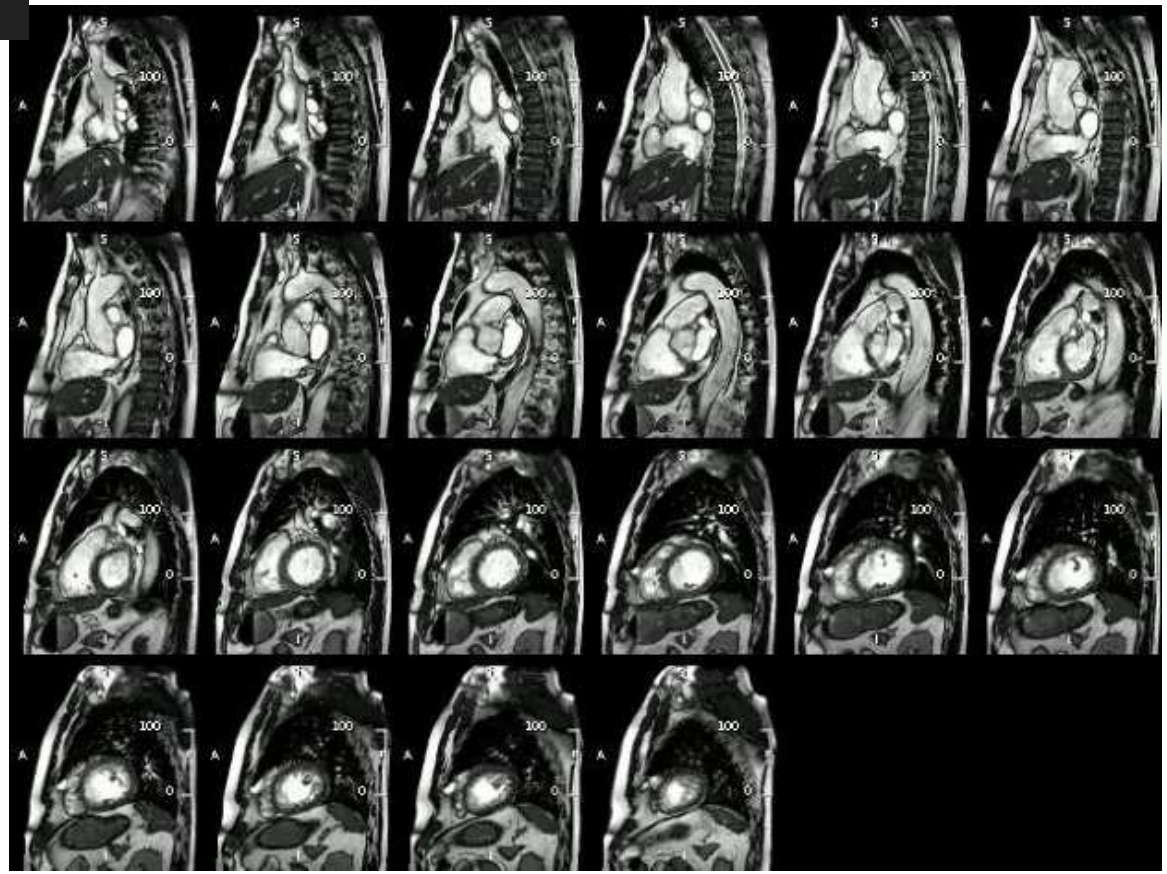


dünne Schichten
lückenlos

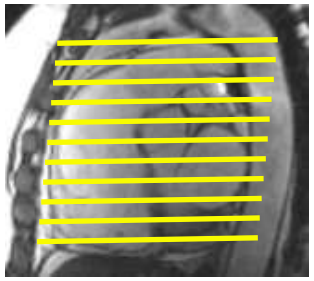


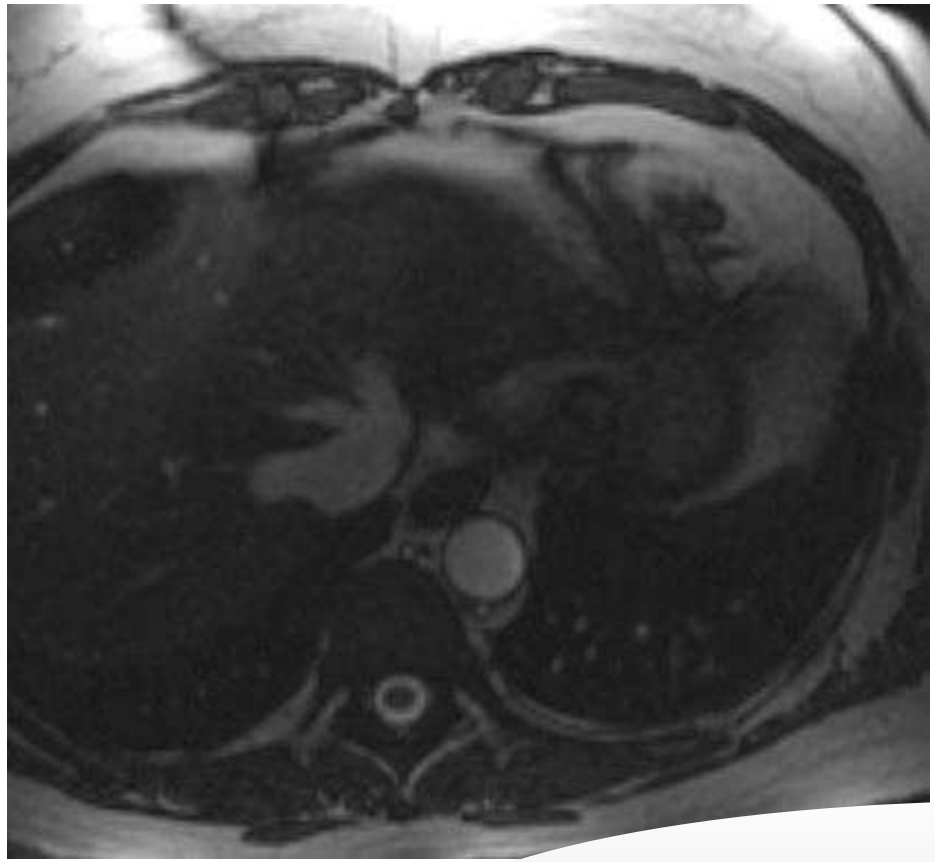
dünne Schichten
lückenlos





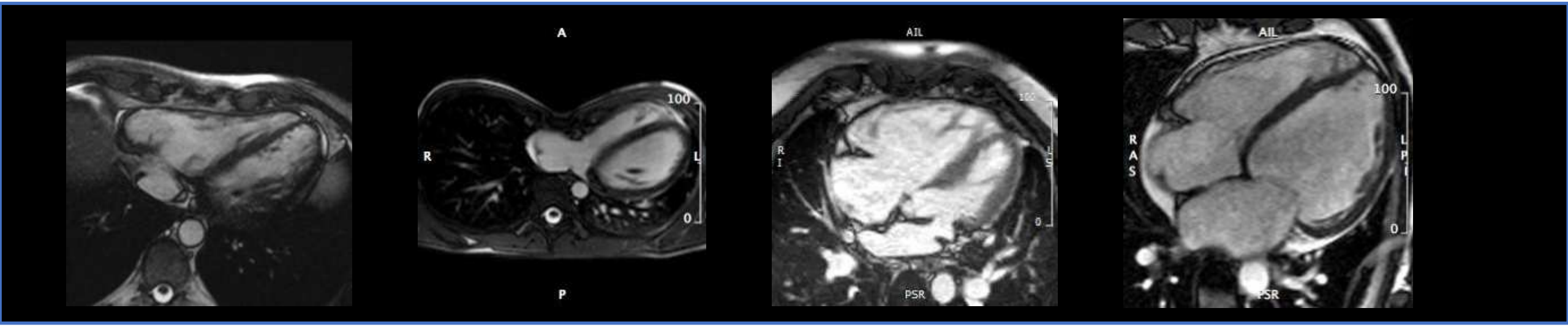
Wandbewegungsstörungen des Rechten Ventrikels





Diagnosestellung AC
NICHT allein aus der Bildgebung!

Rechter Ventrikel - Challenge für die Bildgebung



Bedeutung einer Trabekularisierung??

Kardiomyopathie mit excessiver Trabekularisierung

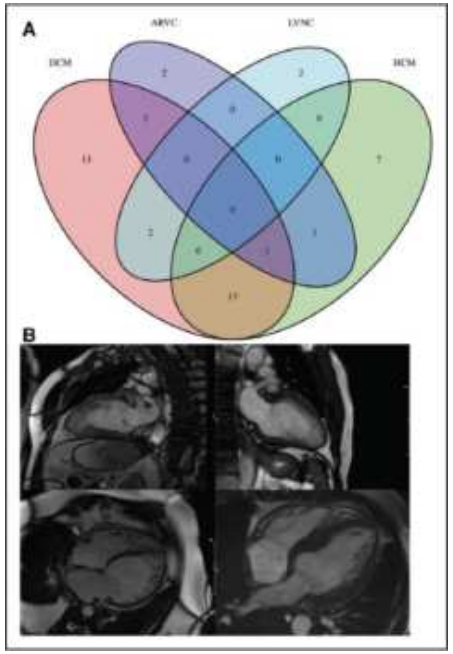
Circulation: Cardiovascular Imaging

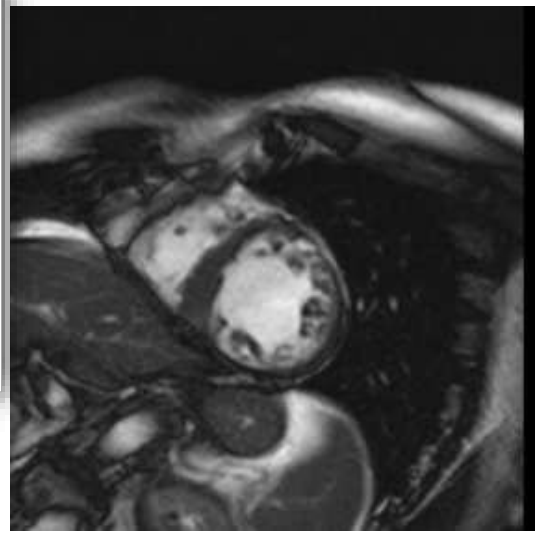
ORIGINAL ARTICLE

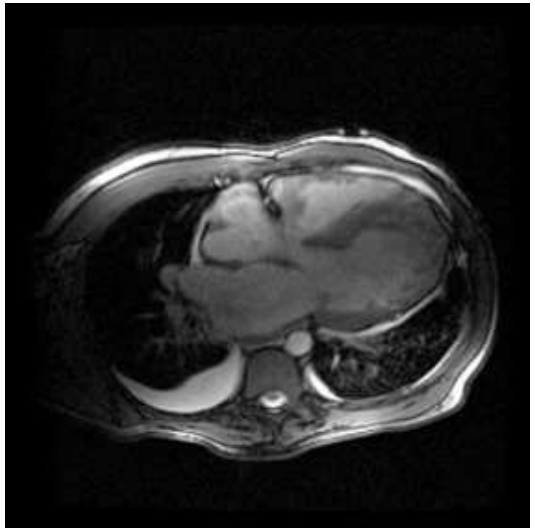
Prognostic Significance of Left Ventricular Noncompaction

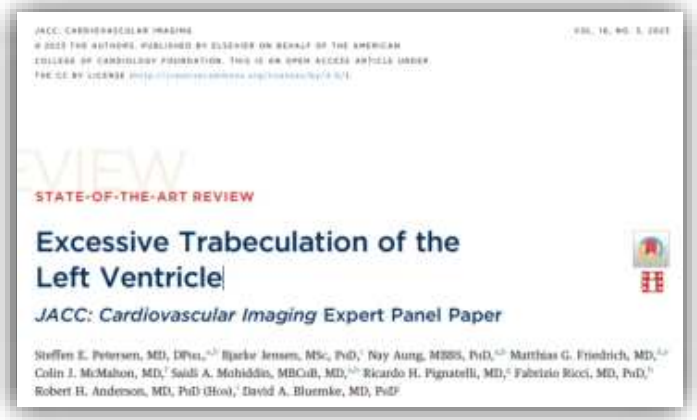
Systematic Review and Meta-Analysis of Observational Studies

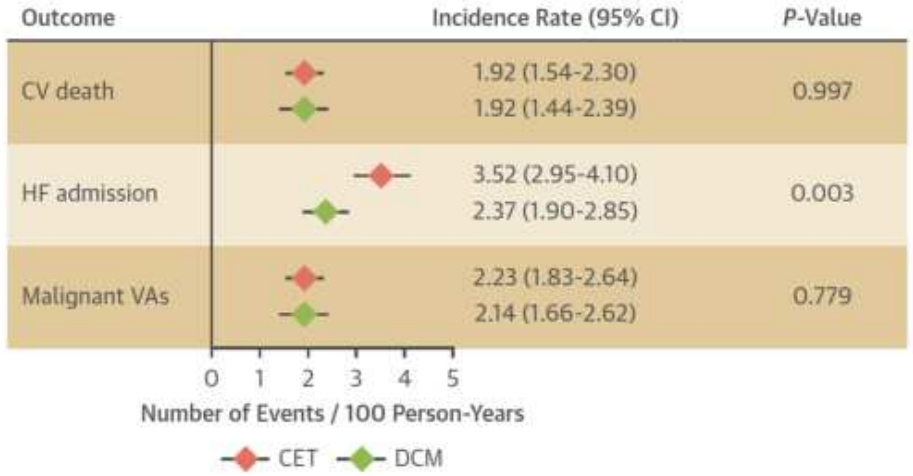
Aung … Petersen 2020





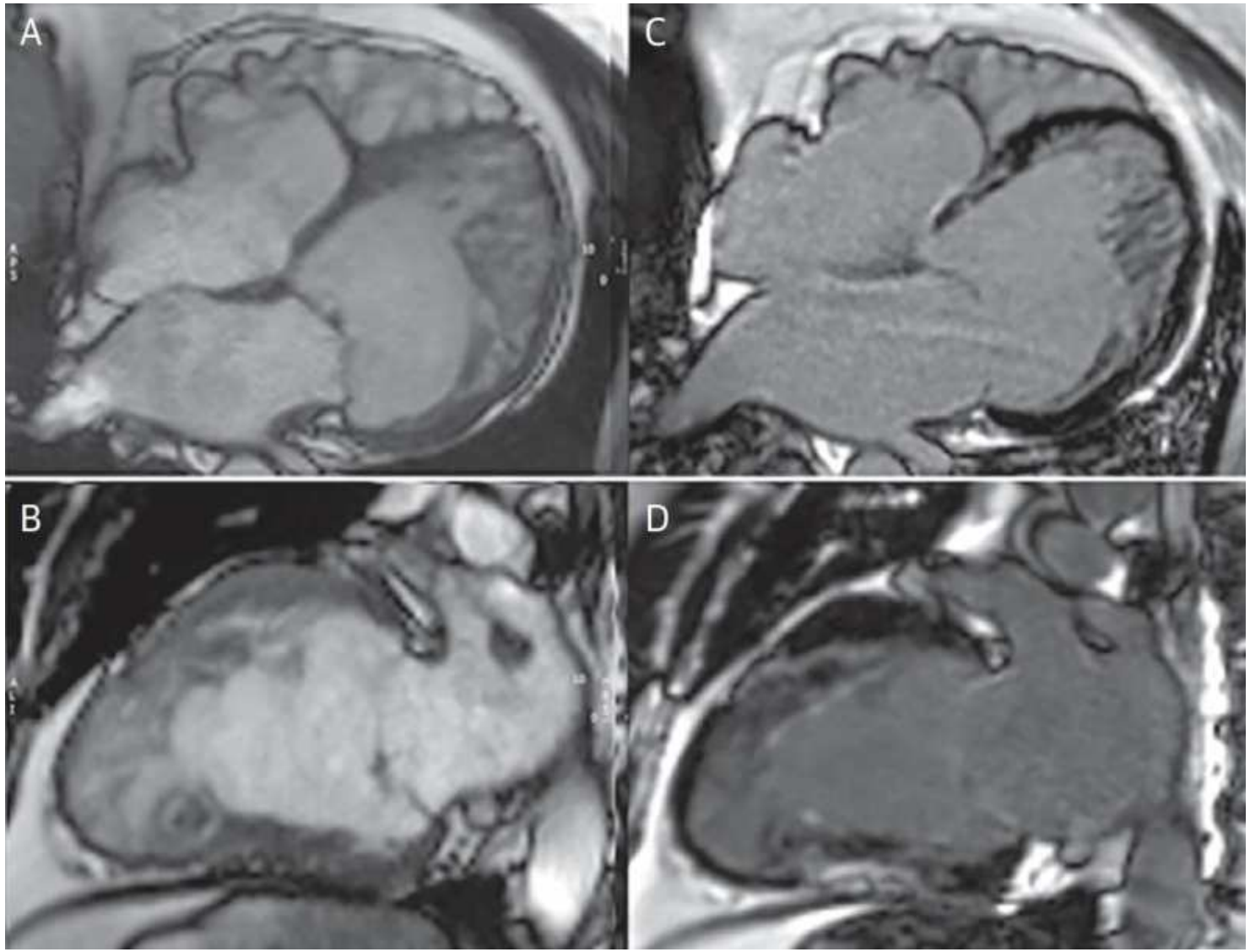






Risiko entspricht DCM

und jetzt?



Petersen et al JACC cvi 2023

European Heart Journal - Cardiovascular Imaging (2017) 18, 237–253
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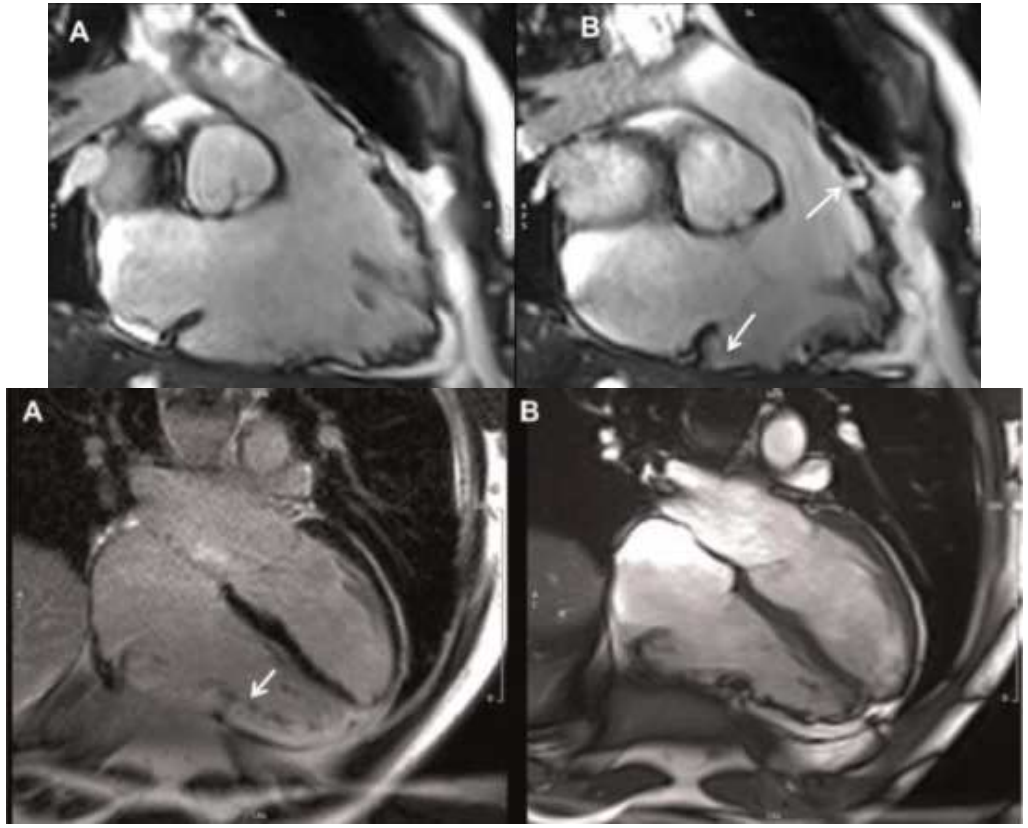
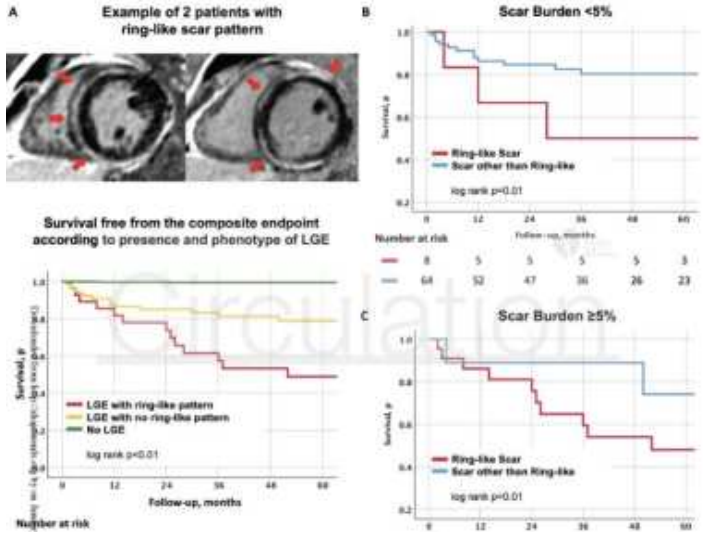
Comprehensive multi-modality imaging approach in arrhythmogenic cardiomyopathy—an expert consensus document of the European Association of Cardiovascular Imaging

Kristina H. Haugaa[1]*, Cristina Basso[2], Luigi P. Badano[3], Chiara Bucciarelli-Ducci[4], Nuno Cardim[5], Oliver Gaemperli[6], Maurizio Galderisi[7], Gilbert Habib[8], Juhani Knuuti[9], Patrizio Lancellotti[10], William McKenna[11], Danilo Neglia[12], Bogdan A. Popescu[13], Thor Edvardsen[1]



- Wandbewegung und Funktion (TF 2010)
- Fokale Fibrose (LGE) : LV-Form?

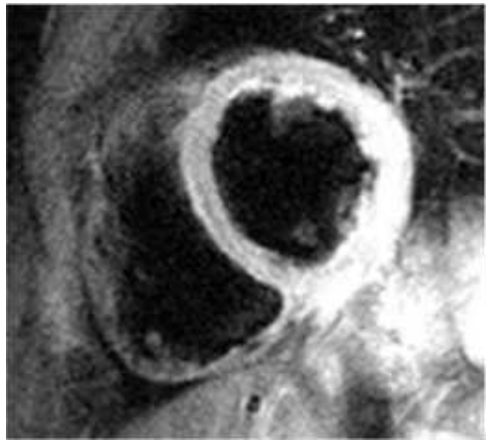
aber - auch bei anderen Erkrankungen

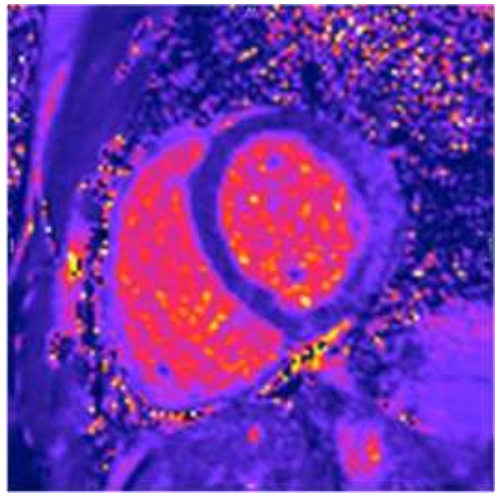


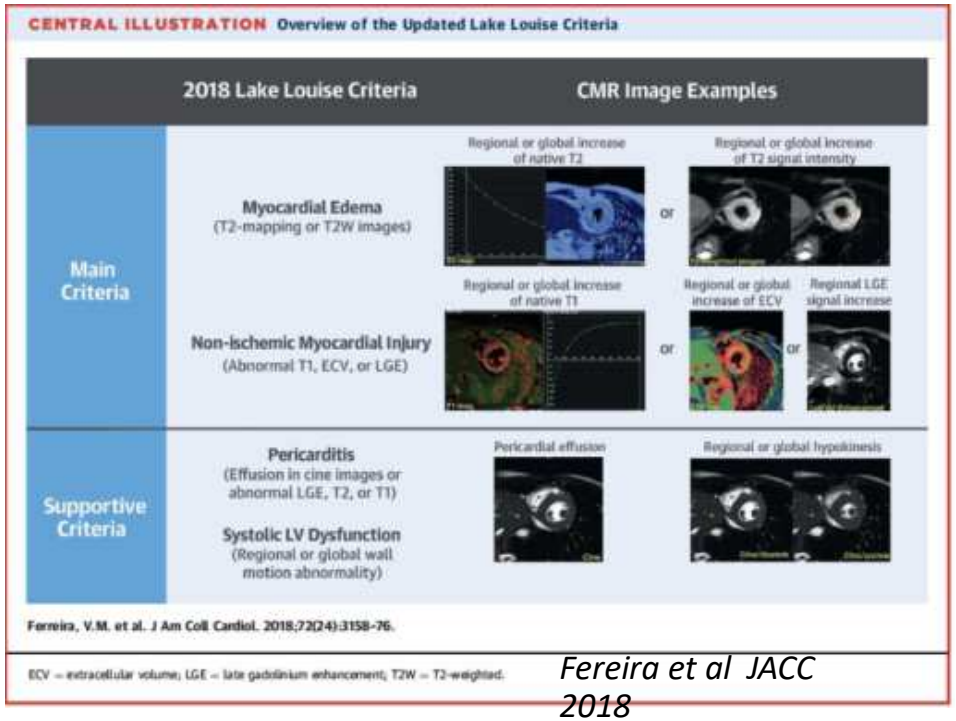
- Kinder und Jugendliche - frühe LV-Beteiligung
  Chungsomprasong et al AJC 2017

Veränderungen beider Ventrikel - Ursachen?

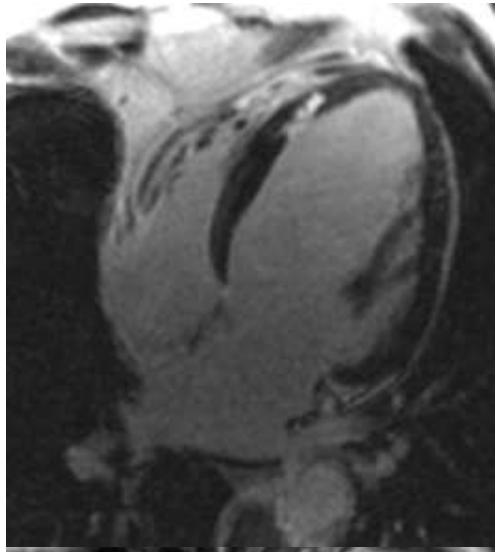
Myokarditis?

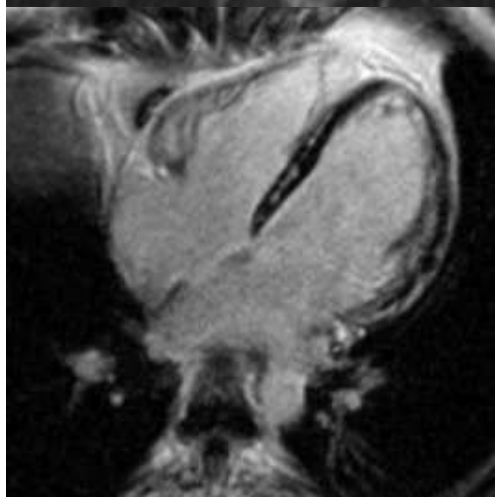




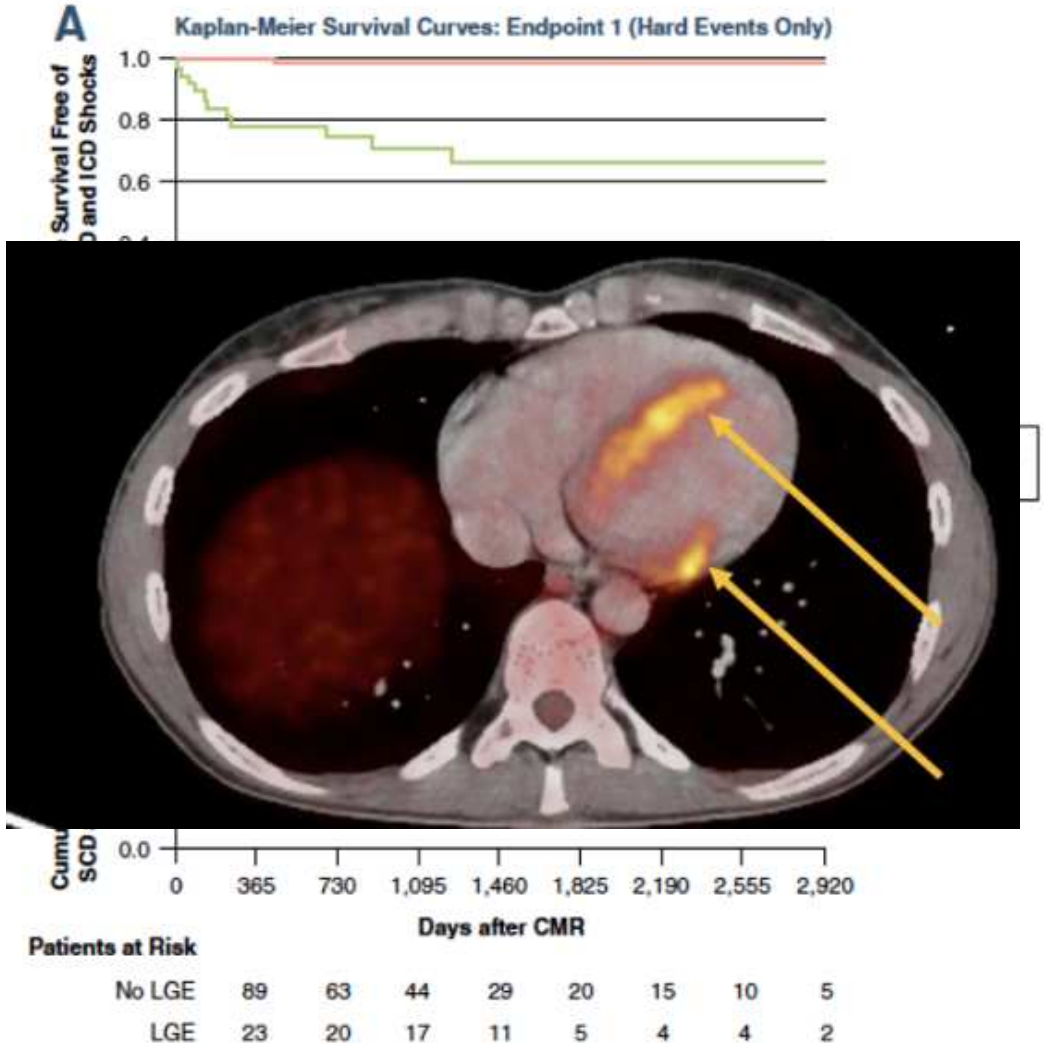


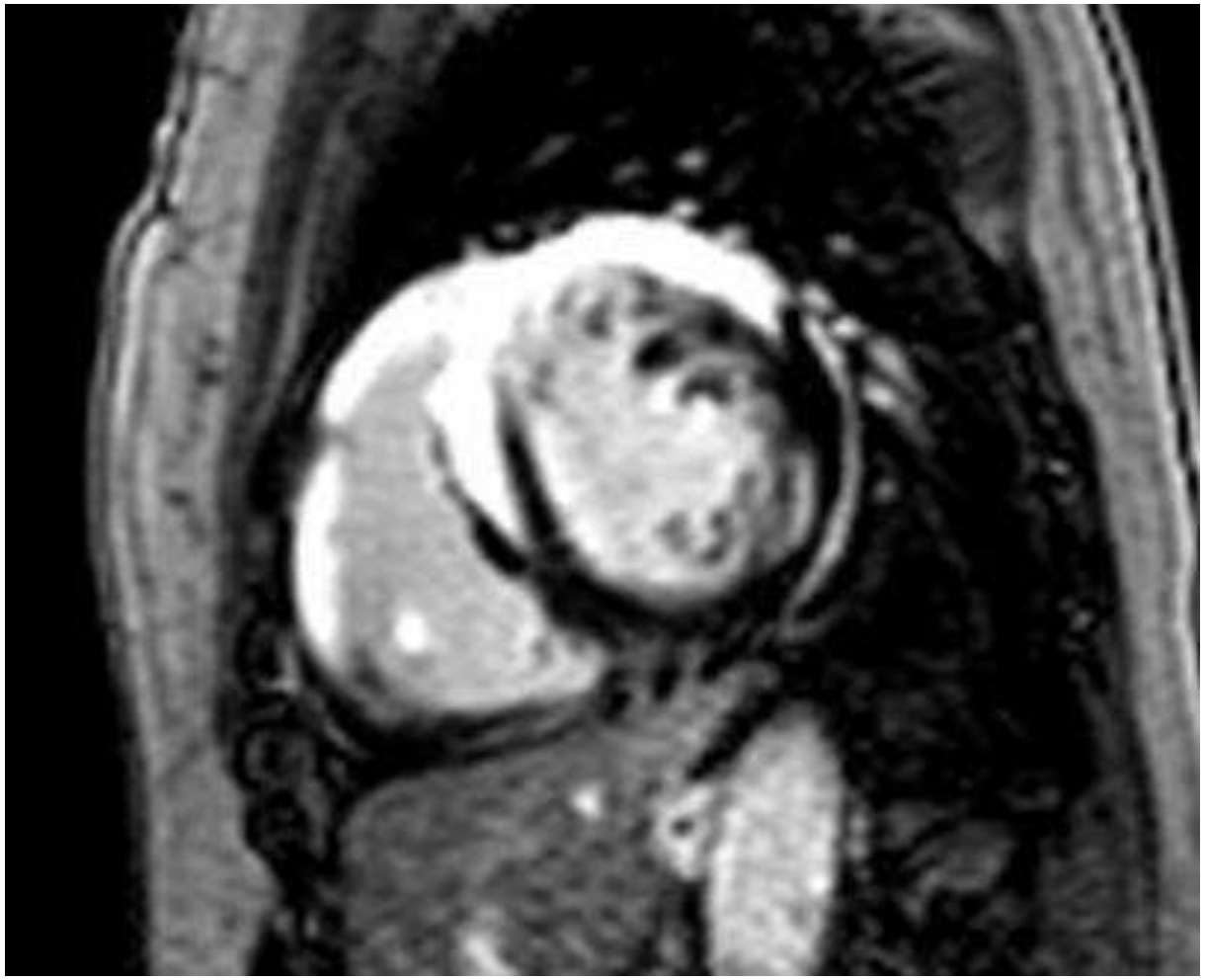
Fereira et al JACC 2018





Sarkoidose - Phänotyp ARVC





Greulich et JACC cvi 2013

International Journal of Cardiology 374 (2023) 120–126

Contents lists available at ScienceDirect

International Journal of Cardiology

journal homepage: www.elsevier.com/locate/ijcard

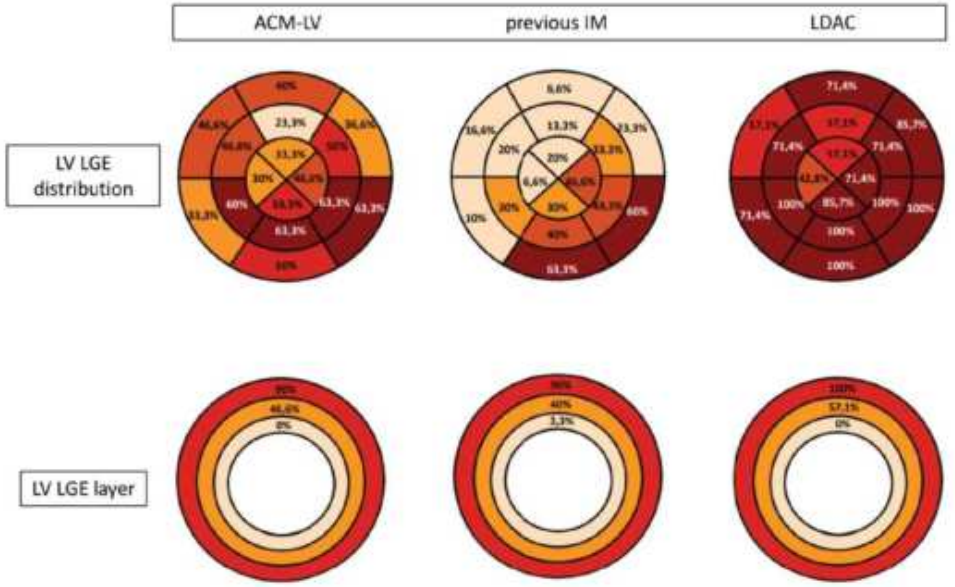
Role of cardiac magnetic resonance in the differential diagnosis between arrhythmogenic cardiomyopathy with left ventricular involvement and previous infectious myocarditis.

Davide Colombo [a,*], Annalisa Turco [b], Sara Lomi [a], Adele Valentini [c], Emilio Bassi [c], Laura Scelsi [b], Alessandra Greco [b], Sandra Schirinzi [b], Michela Zacchino [c], Mauro Acquaro [a], Federico Oliveri [a], Lorenzo Preda [c,d], Stefano Ghio [b]

[a] Department of Molecular Medicine, University of Pavia, Pavia, Italy
[b] Division of Cardiology, Fondazione IRCCS Policlinico San Matteo, Pavia, Italy
[c] Division of Radiology, Fondazione IRCCS Policlinico San Matteo, Pavia, Italy
[d] Department of Clinical, Surgical, Diagnostic, and Pediatric Sciences, University of Pavia, Pavia, Italy

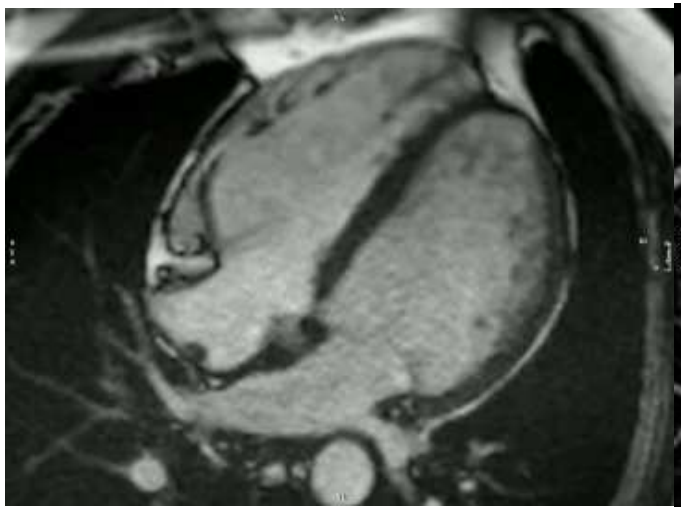
Vorsicht ungeeignet für Routine

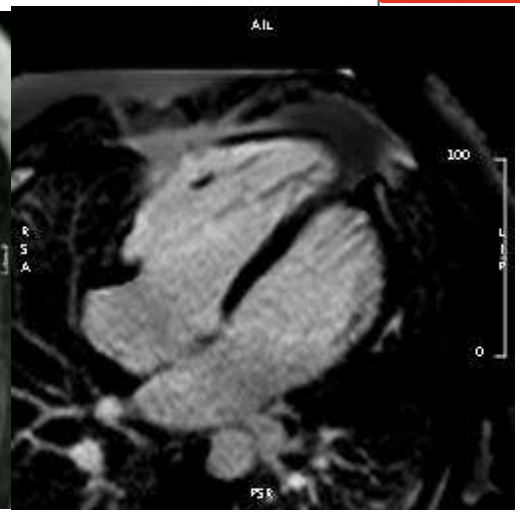
Suggestive finds for LDAC
LGE >15 g
LGE >30%
LV reduced thicknesses
LV wall motion anomalies
septal LGE



Myokardschäden des Rechten Ventrikels - Narben

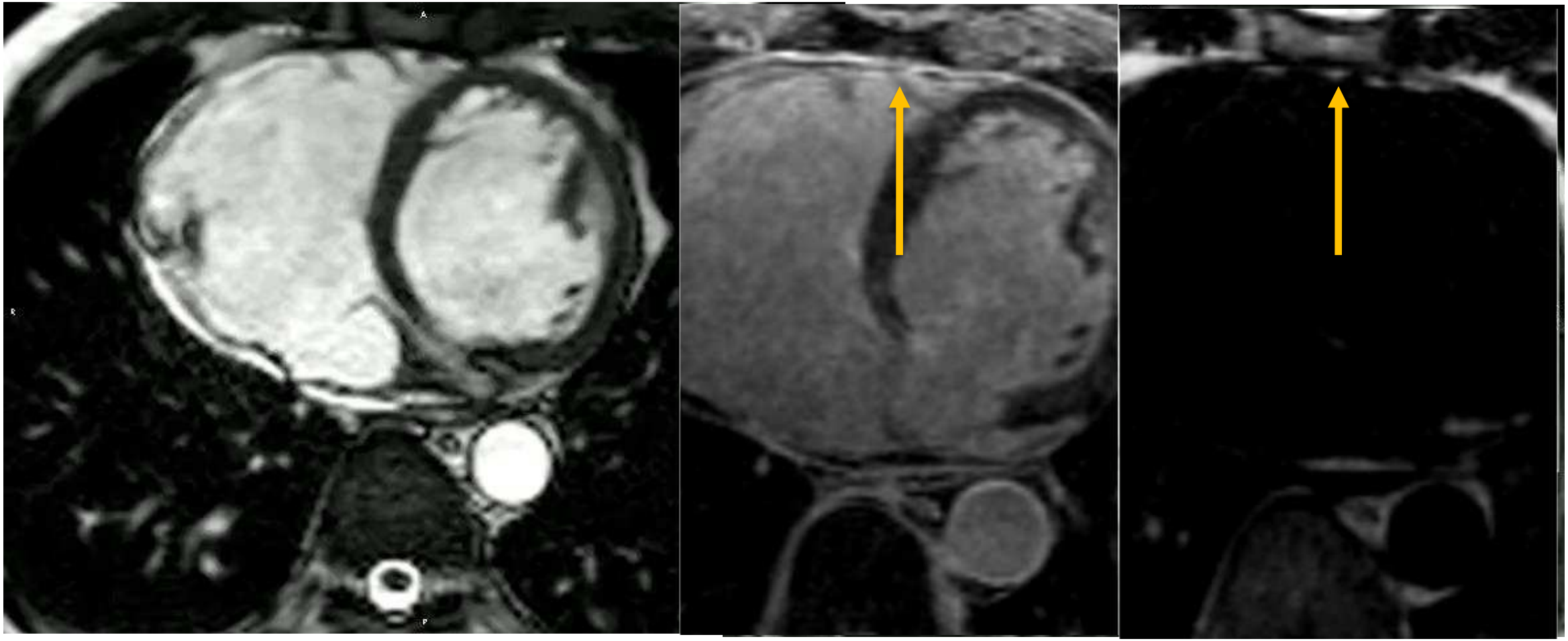
II. Tissue characterization	*Major* *By EMB* • Residual myocytes <60% by morphometric analysis (or 50% if estimated), with fibrous replacement of the RV free wall myocardium in ≥1 sample, with or without fatty replacement of tissue on endomyocardial biopsy *Minor* *By EMB* • Residual myocytes 60% to 75% by morphometric analysis (or 50–65% if estimated), with fibrous replacement of the RV free wall myocardium in ≥1 sample, with or without fatty replacement of tissue on endomyocardial biopsy	*Major* *By CE-CMR:* • Transmural LGE (stria pattern) of ≥1 RV region(s) (inlet, outlet, and apex in 2 orthogonal views) *Major* *By EMB (limited indications):* • Fibrous replacement of the myocardium in ≥1 sample, with or without fatty tissue





RV-Wand Myokardschäden

Zukunft ? Unterscheidung Fett und Fibrose



Prototyp 3D whole heart developed in Koop Siemens und KCL

Diagnose von ARVC mit Schwerpunkt MRT

1. Die Diagnose und Risikostratifizierung bedarf vieler Bausteine
2. Differentialdiagnosen abklären

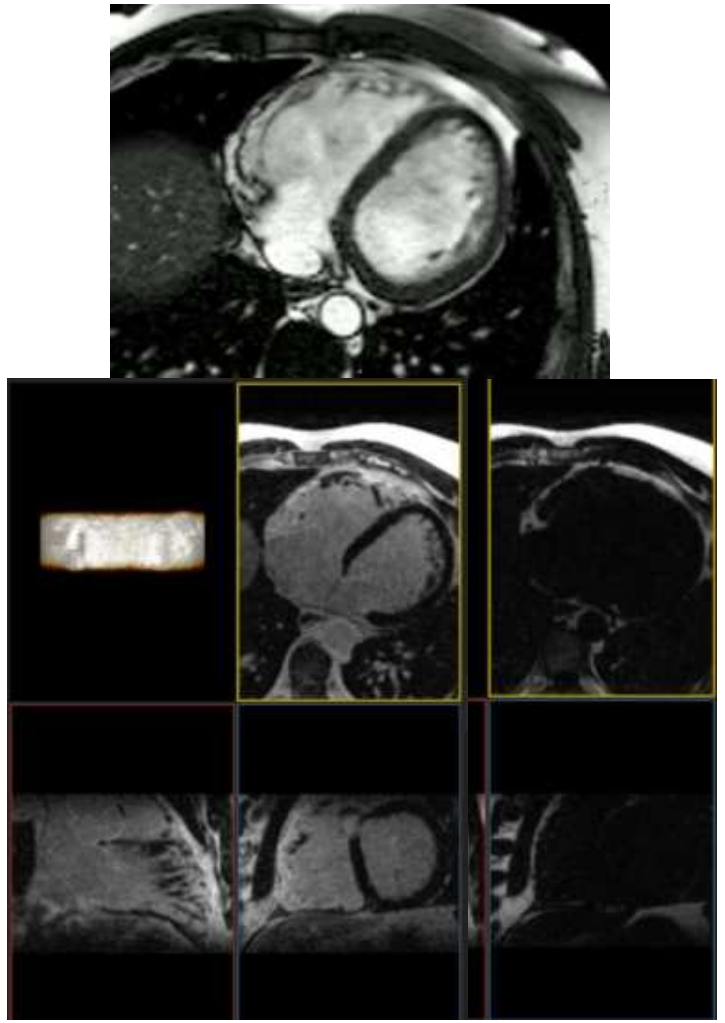
Bildgebung

3. Quantitative und qualitative Beurteilung beider Ventrikel

4. Nachweis von Narben und Fibrose insbesondere linker Ventrikel

5. Herausforderung: Narben und Fibrosen rechter Ventrikel

6. Sicherer Nachweis von Fettinfiltrationen in (naher) Zukunft



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