

Arytmogenní kardiomyopatie pravé komory

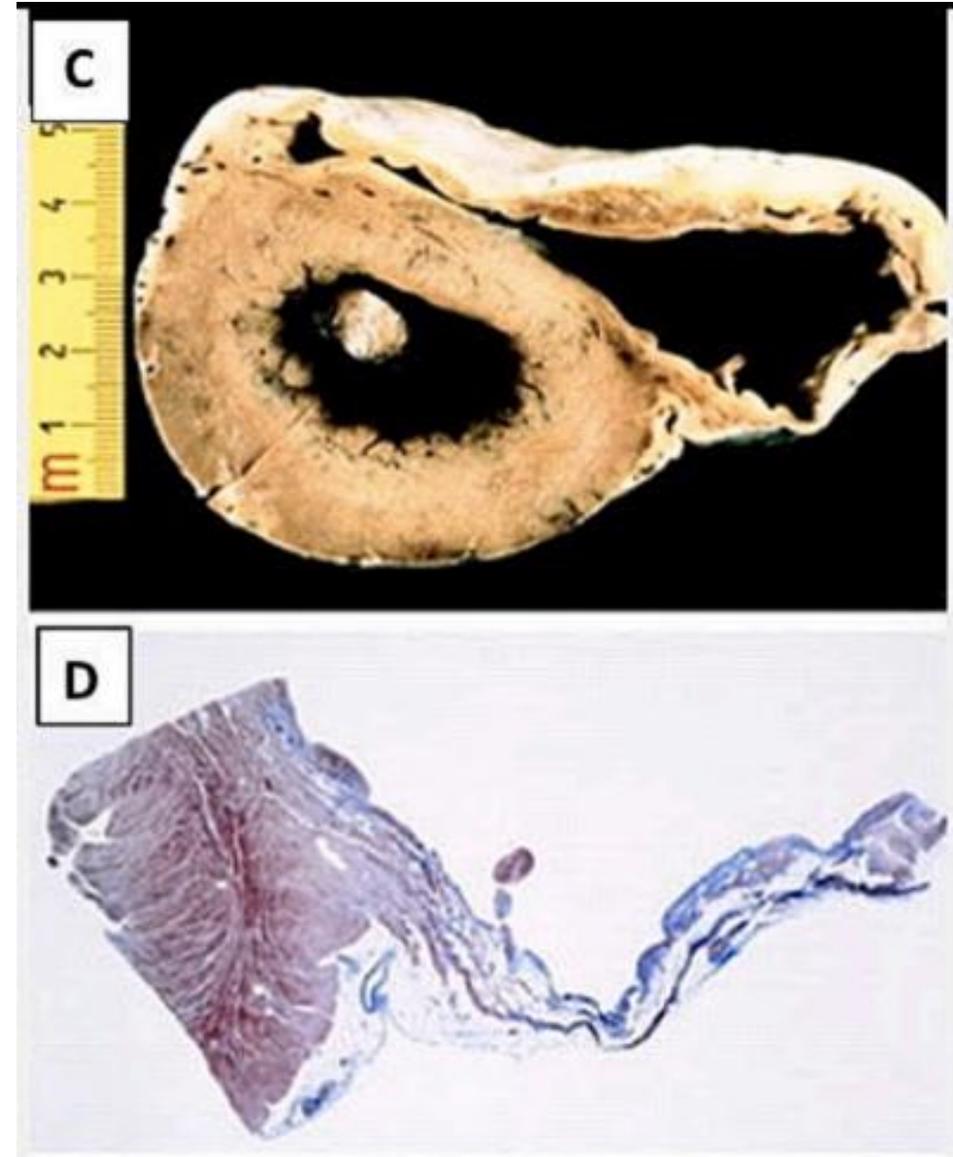
využití multimodalitního zobrazování pro stratifikaci rizika u pacientů
s komorovými arytmiemi

Pleva Martin

Nemocnice Podlesí, a.s., Třinec
Vítkovická nemocnice, a.s., Ostrava

Arytmogenní KMP

- geneticky podmíněné onemocnění
- tukově-fibrózní náhrada myokardu komor
- riziko komorových dysrytmií...srdečního selhání



Prevalence a riziko náhlé srdeční smrti

Cardiomyopathy phenotype	Adults	Children
HCM	Prevalence: 0.2% ²⁶⁻³³	Childhood incidence: 0.002–0.005% ³⁴⁻³⁶ Childhood prevalence: 0.029% ³⁶
DCM	Prevalence: 0.036–0.400% ^{25,37}	Childhood incidence: 0.003–0.006% Childhood prevalence: 0.026% ³⁶ Infantile incidence: 0.038–0.046% ^{34-36,38}
NDLVC	To be determined	To be determined
ARVC	Prevalence: 0.078% ³⁹⁻⁴¹	Very rare in infancy and early childhood; to be determined in older children and adolescents
RCM	Rare	Childhood incidence: 0.0003% ³⁴

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výskyt VT/VF, adekv. výboje ICD, NSS:

- ARVC – 10,6 %/rok
- G+/P- – 3,7 %/rok

Meta-Analysis > Heart Rhythm. 2018 Jul;15(7):1097-1107. doi: 10.1016/j.hrthm.2018.01.031. Epub 2018 Feb 3.

Predicting arrhythmic risk in arrhythmogenic right ventricular cardiomyopathy: A systematic review and meta-analysis

Laurens P Bosman¹, Arjan Sammani², Cynthia A James³, Julia Cadrin-Tourigny⁴, Hugh Calkins⁵, J Peter van Tintelen⁶, Richard N W Hauer⁷, Folkert W Asselbergs⁸, Anneline S J M Te Riele⁹

Affiliations + expand

PMID: 29408436 DOI: 10.1016/j.hrthm.2018.01.031



Diagnosis of arrhythmogenic cardiomyopathy: The Padua criteria

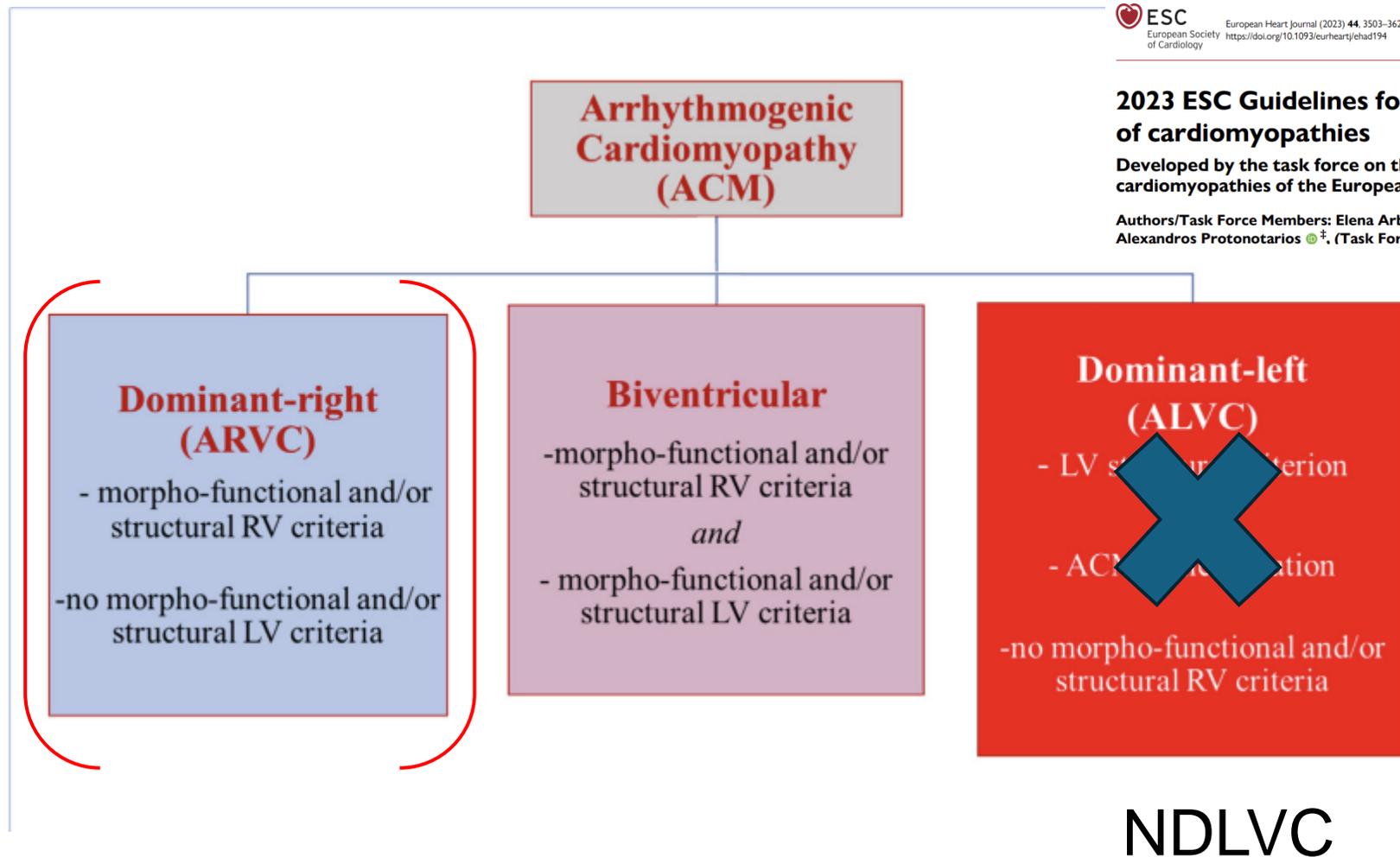
Domenico Corrado ^{a,*}, Martina Perazzolo Marra ^a, Alessandro Zorzi ^a, Giorgia Boffagna ^a, Alberto Cipriani ^a, Manuel De Lazzari ^a, Federico Migliore ^a, Kalliopi Pilichou ^a, Alessandra Rampazzo ^b, Ilaria Rigato ^a, Stefania Rizzo ^a, Gaetano Thiene ^a, Aris Anastasakis ^c, Angeliki Asimaki ^d, Chiara Bucciarelli-Ducci ^e, Kristine H. Haugaa ^f, Francis E. Marchlinski ^g, Andrea Mazzanti ^h, William J. McKenna ⁱ, Antonis Pantazis ^j, Antonio Pelliccia ^k, Christian Schmied ^l, Sanjay Sharma ^m, Thomas Wichter ⁿ, Barbara Bauce ^a, Cristina Basso ^a

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).

AKMP - fenotypy



Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia

Proposed Modification of the Task Force Criteria

Frank I. Marcus^{1a}, Chair, William J. McKenna² Co-Chair, Duane Sherrill¹, Cristina Basso³, Barbara Baucé³, David A. Bluemke⁴, Hugh Calkins⁵, Domenico Corrado³, Moniek G.P.J. Cox⁴, James P. Daubert⁷, Guy Fontaine¹⁰, Kathleen Gear¹, Richard Hauer⁴, Andrea Nava⁷, Michael H. Picard¹¹, Nikos Protonotarios¹³, Jeffrey E. Saffitz¹², Danita M. Yoerger Sanborn¹¹, Jonathan S. Steinberg⁹, Harikrishna Tandri¹³, Gaetano Thiene⁴, Jeffrey A. Towbin¹⁴, Adalena Tsatsopoulou¹³, Thomas Wichter¹⁵, and Wojciech Zareba⁸

cká

Diagnosis of arrhythmogenic cardiomyopathy: The Padua criteria

Domenico Corrado^{a,b}, Martina Perazzolo Marra^a, Alessandro Zorzi^a, Giorgia Beffagna^a, Alberto Cipriani^a, Manuel De Lazzari^a, Federico Migliore^a, Kalliopi Pilichou^a, Alessandra Rampazzo^b, Ilaria Rigato^a, Stefania Rizzo^a, Gaetano Thiene^a, Aris Anastasakis^c, Angeliki Asimaki^d, Chiara Bucciarelli-Ducci^e, Kristine H. Haugaa^f, Francis E. Marchlinski^g, Andrea Mazzanti^h, William J. McKennaⁱ, Antonis Pantazis^j, Antonio Pelliccia^k, Christian Schmied^l, Sanjay Sharma^m, Thomas Wichterⁿ, Barbara Baucé^a, Cristina Basso^a

Criteria for diagnosis of right ventricular dysplasia

I Global and/or regional dysfunction and structural alterations^{17–23} *

MAJOR

Severe dilatation and reduction of right ventricular ejection fraction with no (or only mild) LV impairment
Localised right ventricular aneurysms (akinetic or dyskinetic areas with diastolic bulging)
Severe segmental dilatation of the right ventricle

MINOR

Mild global right ventricular dilatation and/or ejection fraction reduction with normal left ventricle
Mild segmental dilatation of the right ventricle
Regional right ventricular hypokinesia

II Tissue characterisation of walls

MAJOR

Fibrofatty replacement of myocardium on endomyocardial biopsy

VI. Family history/genetics

III Repolarisation abnormality

MINOR

Inverted T waves in right precordial leads (V2 (people aged more than 12 yr; in absence of right bundle branch block)

IV Depolarisation/conduction abnormalities

MAJOR

Epsilon waves or localised prolongation (>110 ms) of the QRS complex in right precordial leads (V1–V3)

MINOR

Late potentials (signal averaged ECG)

V Arrhythmias

MINOR

Left bundle branch block type ventricular tachycardia (sustained and non-sustained) (ECG, Holter, exercise testing).
Frequent ventricular extrasystoles (more than 1000/24 h) (Holter)

Major

- Identification of a pathogenic ACM-gene variant in the patient under evaluation
- ACM confirmed in a first-degree relative who meets diagnostic criteria
- ACM confirmed pathologically at autopsy or surgery in a first-degree relative

Minor

- Identification of a likely-pathogenic ACM-gene variant in the patient under evaluation
- History of ACM in a first-degree relative in whom it is not possible or practical to determine whether the family member meets diagnostic criteria
- Premature sudden death (<35 years of age) due to suspected ACM in a first-degree relative
- ACM confirmed pathologically or by diagnostic criteria in second-degree relative

*Detected by echocardiography, angiography, magnetic resonance imaging, or radionuclide scintigraphy. ECG, electrocardiogram; LV, left ventricle.

Diagnostická kritéria

Diagnóza potvrzena (při splnění):

- 2 velkých kritérií
- 1 velkého a dvou malých kritérií
- 4 malých kritérií

„Hraniční“ diagnóza:

- 1 velké a 1 malé kritérium
- 3 malá kritéria

„Možná“ diagnóza:

- 1 velké nebo 2 malá kritéria

Diagnostická kritéria 2024

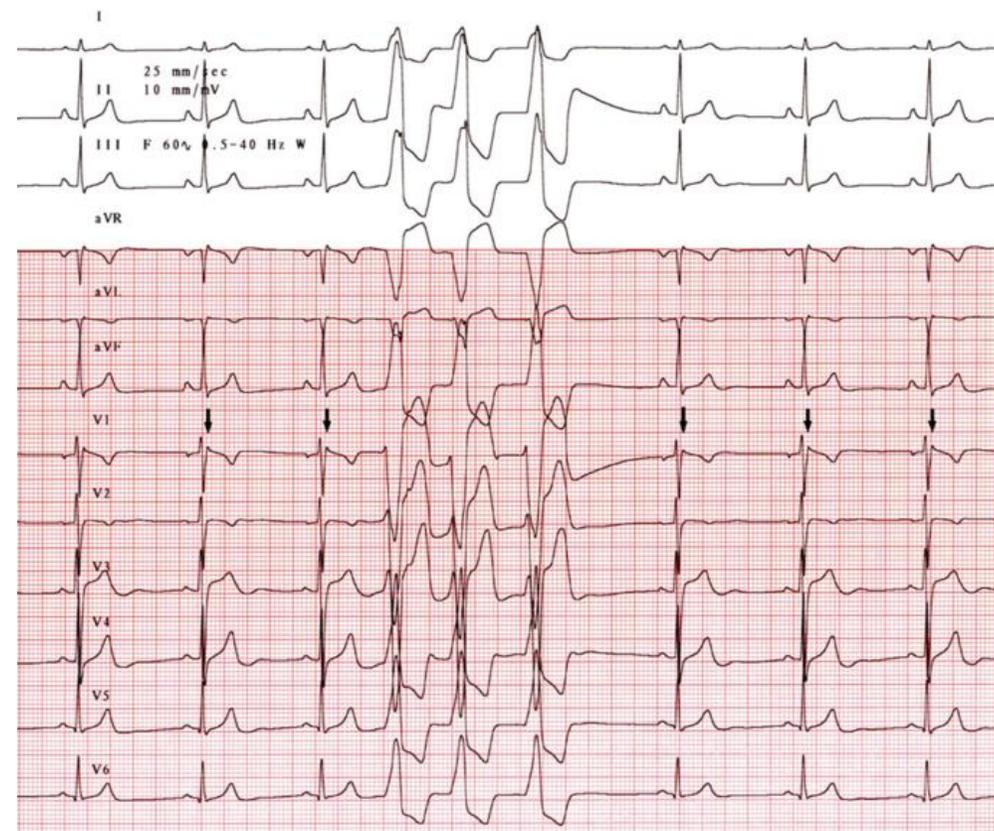
V. Arrhythmias

Major

- Frequent ventricular extrasystoles (>500 per 24 h), non-sustained or sustained ventricular tachycardia of LBBB morphology with non-inferior axis

Minor

- Frequent ventricular extrasystoles (>500 per 24 h), non-sustained or sustained ventricular tachycardia of LBBB morphology with inferior axis (“RVOT pattern”)
- History of cardiac arrest due to ventricular fibrillation or sustained ventricular tachycardia of unknown morphology



https://link.springer.com/chapter/10.1007/978-3-030-38662-7_5

Diagnostická kritéria 2024

VI. Family history/genetics

III. Repolarization abnormalities

Major

- Negative T waves in right precordial leads (V₁, V₂, and V₃) or beyond in individuals ≥ 14 year-old (in the absence of complete RBBB and not preceded by J-point/ST-segment elevation)

Minor

- Negative T waves in leads V₁ and V₂ in males ≥ 14 year-old (in the absence of RBBB and not preceded by J-point/ST-segment elevation)
- Negative T waves beyond V₃ in the presence of complete RBBB
- Negative T waves beyond V₃ in individuals < 14 year-old

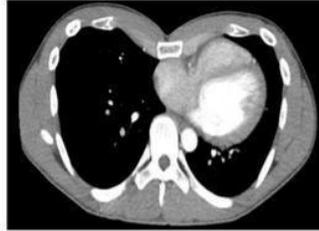
IV. Depolarization and conduction abnormalities

Minor

- Epsilon wave (reproducible low-amplitude signals between end of QRS complex to onset of the T wave) in the right precordial leads (V₁ to V₃)
- Terminal activation duration of QRS ≥ 55 ms measured from the nadir of the S wave to the end of the QRS, including R', in V₁, V₂, or V₃ (in the absence of complete RBBB)

Specificita EKG kritérií?

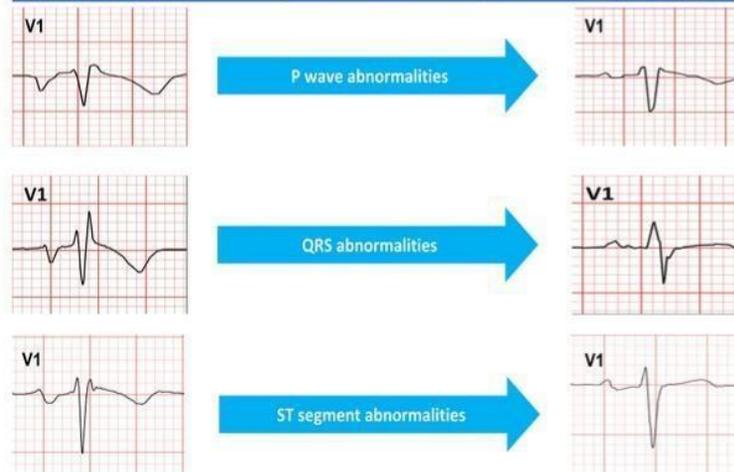
The electrocardiographic manifestations of pectus excavatum before and after surgical correction



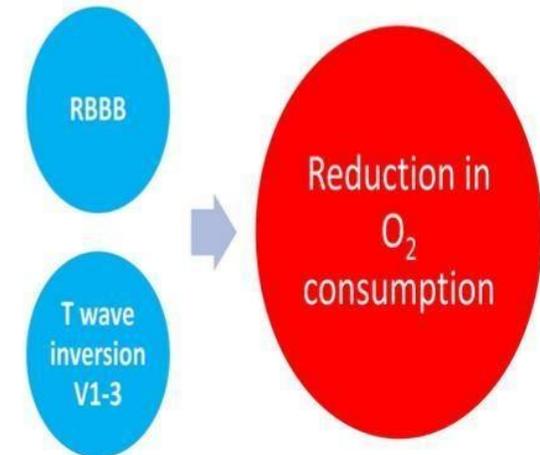
Pectus excavatum patients who completed a **minimally invasive surgical repair** and had a **preoperative and a postoperative ECG evaluation**

310 patients
between
2010 and
2022

Most common ECG changes before and after pectus excavatum repair



Association between ECG abnormalities and O₂ consumption



Electrocardiographic abnormalities in PEx are common and can revert to normal following surgical repair. Preoperative RBBB and T wave inversion in leads V1-3 suggested a reduction in exercise capacity, serving as a marker for the need for further cardiovascular evaluation of these patients.

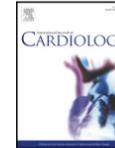


Journal of Electrocardiology
Volume 82, January–February 2024, Pages 19–26



The electrocardiographic manifestations of pectus excavatum before and after surgical correction

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Olubadewa A. Fatunde MD MPH^b, Dan Sorajja MD^b



Diagnosis of arrhythmogenic cardiomyopathy: The Padua criteria



Domenico Corrado ^{a,*}, Martina Perazzolo Marra ^a, Alessandro Zorzi ^a, Giorgia Beffagna ^a, Alberto Cipriani ^a, Manuel De Lazzari ^a, Federico Migliore ^a, Kalliopi Pilichou ^a, Alessandra Rampazzo ^b, Ilaria Rigato ^a, Stefania Rizzo ^a, Gaetano Thiene ^a, Aris Anastasakis ^c, Angeliki Asimaki ^d, Chiara Bucciarelli-Ducci ^e, Kristine H. Haugaa ^f, Francis E. Marchlinski ^g, Andrea Mazzanti ^h, William J. McKenna ⁱ, Antonis Pantazis ^j, Antonio Pelliccia ^k, Christian Schmied ^l, Sanjay Sharma ^m, Thomas Wichter ⁿ, Barbara Bauce ^a, Cristina Basso ^a

At variance with genetically-determined cardiac ion channel disorders, pathogenic mutations, ECG abnormalities or arrhythmias are not sufficient for diagnosis of ACM, which is a structural heart muscle disease. By analogy with the diagnosis of hypertrophic cardiomyopathy, that requires demonstration of critical LV hypertrophy [23], at least one RV or LV criterion from categories I (i.e., morpho-functional ventricular abnormalities) or II (i.e., structural myocardial abnormalities), either major or minor is mandatory to reach a diagnosis of ACM according to the Padua criteria (Fig. 2).

Zobrazovací metody

- *Angiografie pravé komory*
- **ECHO**
- **CMR**
- *CT*
- *FDG-PET/CT*
- *Elektroanatomické mapování*

Zobrazovací metody

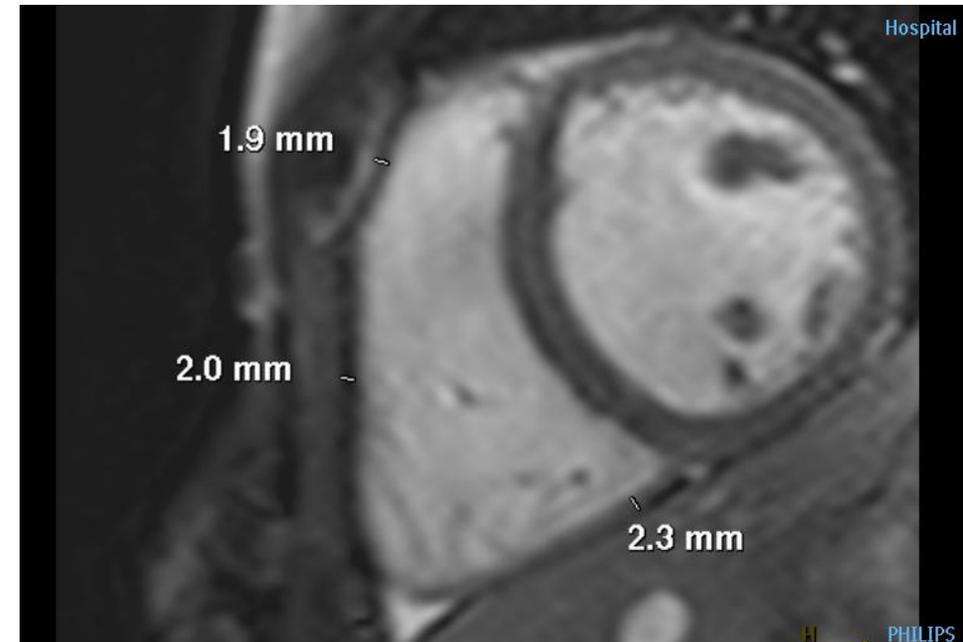
I. Morpho-functional ventricular abnormalities

Major

- Regional RV akinesia, dyskinesia, or aneurysm plus one of the following:
 - 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

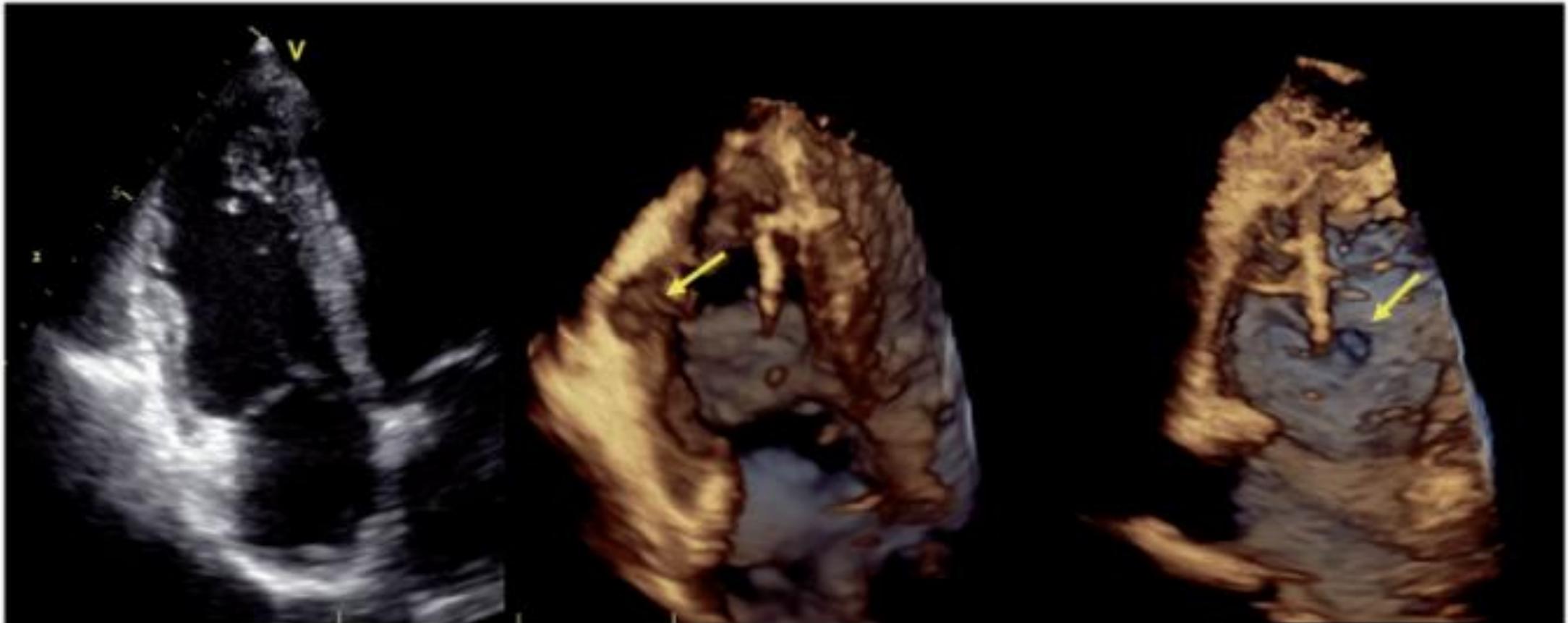
- Regional RV akinesia, dyskinesia or aneurysm of RV free wall



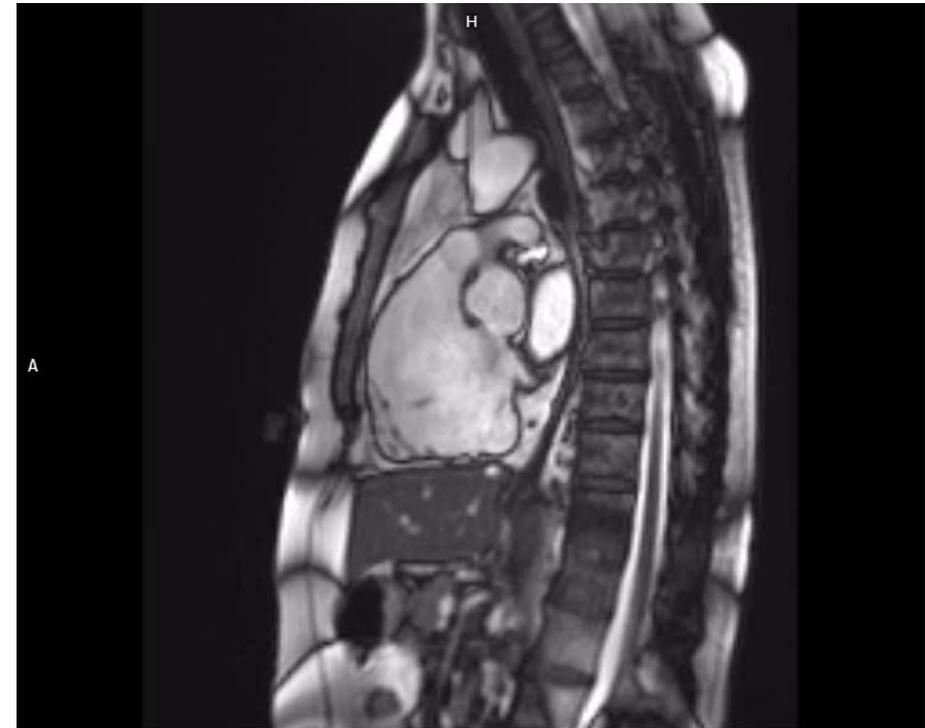
Porucha kinetiky - fokální

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

Kristina H. Haugaa^{1*}, Cristina Basso², Luigi P. Badano³, Chiara Bucciarelli-Ducci⁴,
Nuno Cardim⁵, Oliver Gaemperli⁶, Maurizio Galderisi⁷, Gilbert Habib⁸,
Juhani Knuuti⁹, Patrizio Lancellotti¹⁰, William McKenna¹¹, Danilo Neglia¹²,
Bogdan A. Popescu¹³, Thor Edvardsen¹

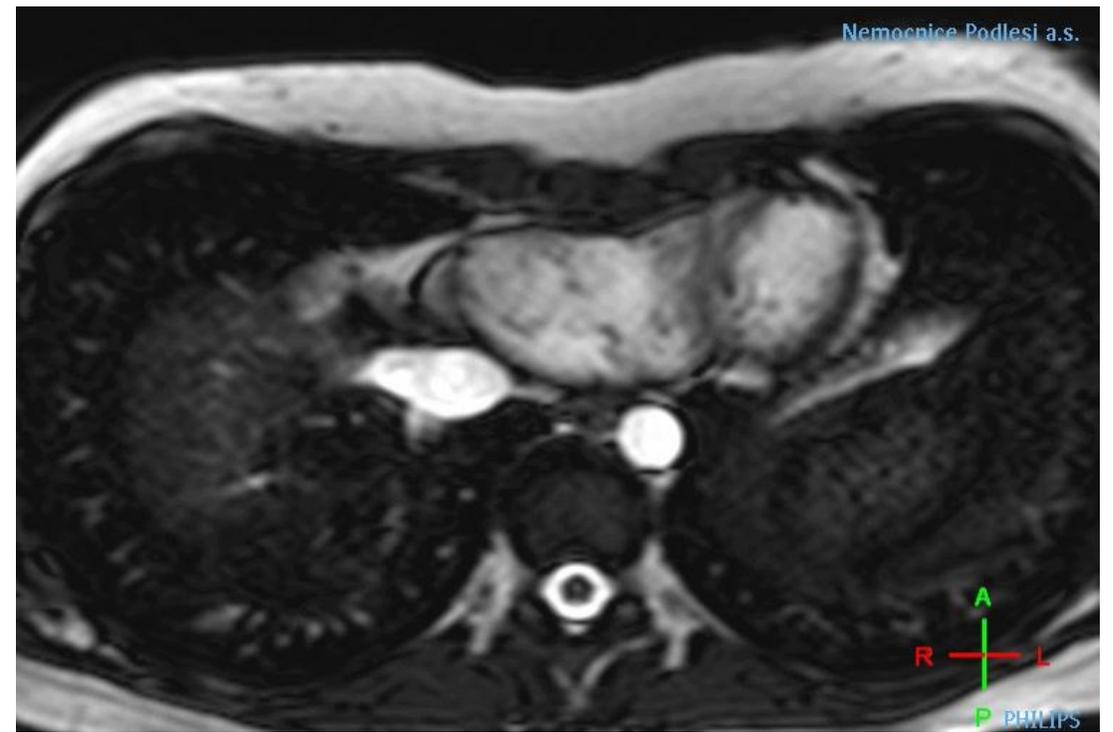
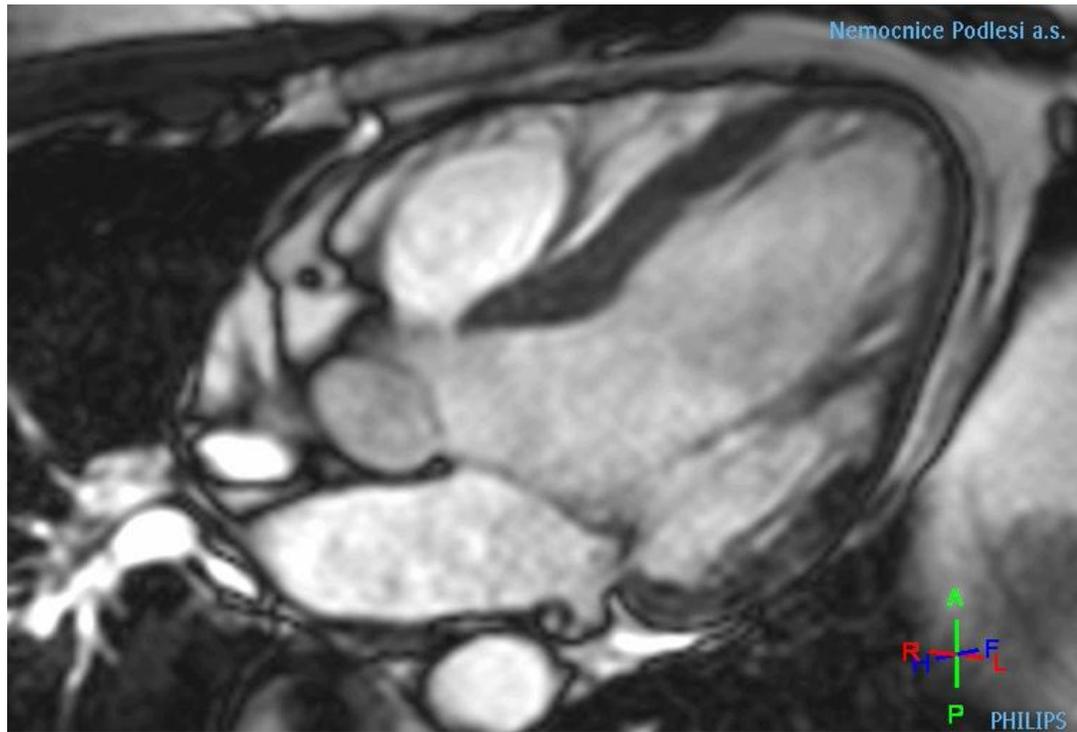


Porucha kinetiky - fokální



Porucha kinetiky - fokální

varianta normy



Zobrazovací metody

I. Morpho-functional ventricular abnormalities

Major

- Regional RV akinesia, dyskinesia, or aneurysm

plus one of the following:

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

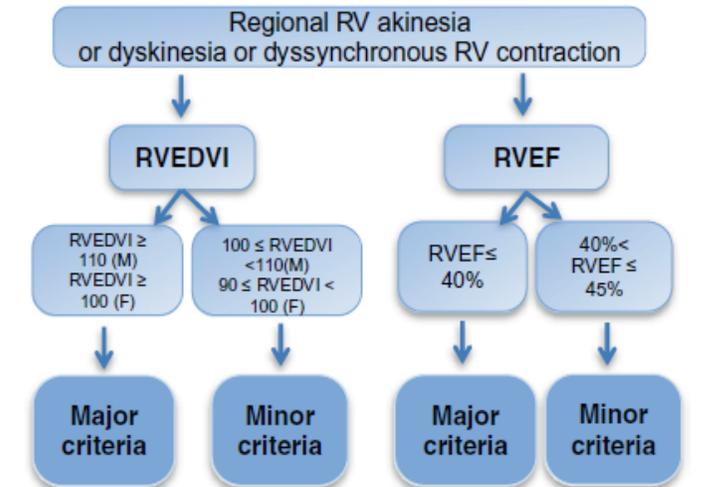
or

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

Minor

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

2010 CMR TFC



allows comparison of the details for 1994 and 2010 ARVC

Liu T et al. Effect of the 2010 task force criteria on reclassification of cardiovascular magnetic resonance criteria for arrhythmogenic right ventricular cardiomyopathy. *Journal of Cardiovascular Magnetic Resonance*. 2014;14(6):1-10.

ECHO

MAJOR ECHOCARDIOGRAPHIC CRITERIA FOR ARVC

Regional RV Dyskinesia or Aneurysm

And one of the following

PLAX RVOT ≥ 32 mm (corrected for body size [PLAX/BSA] ≥ 19 mm/m²)

PSAX RVOT ≥ 36 mm (corrected for body size [PLAX/BSA] ≥ 21 mm/m²)

Or

Fractional Area Change $\leq 33\%$

MINOR ECHOCARDIOGRAPHIC CRITERIA FOR ARVC

Regional RV Akinesia or Dyskinesia

And one of the following

PLAX RVOT ≥ 29 to < 32 mm (corrected for body size [PLAX/BSA] ≥ 16 to < 19 mm/m²)

PSAX RVOT ≥ 32 to < 36 mm (corrected for body size [PLAX/BSA] ≥ 18 to 21 mm/m²)

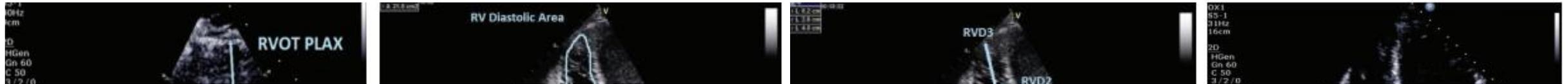
Or

Fractional Area Change > 33 to $< 40\%$

ECHO

Utility of Tissue Doppler and Strain Echocardiography in Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy

Kalpana R. Prakasa, MD, Jianwen Wang, MD, Harikrishna Tandri, MD, Darshan Dalal, MBBS, Chandra Bomma, MD, Roman Chojnowski, RDCS, Cynthia James, PhD, Crystal Tichnell, MGC, Stuart Russell, MD, Daniel Judge, MD, Mary Corretti, MD, David Bluemke, MD, Hugh Calkins, MD, and Theodore P. Abraham, MD*



Diagnostic value of tissue Doppler and strain echocardiography

Parameter	Cut-Off Value	Specificity (%)	Sensitivity (%)	AUC	p Value
RV systolic velocity	7.5 cm/s	89	67	0.83	0.0001
RV displacement	18 mm	71	77	0.77	0.002
RV strain	-18%	87	73	0.82	0.004
RV strain rate	-0.76 s ⁻¹	96	50	0.76	0.01
RV outflow tract diameter	3 cm	94	53	0.73	0.01
RV fractional area change	31%	83	47	0.61	0.26

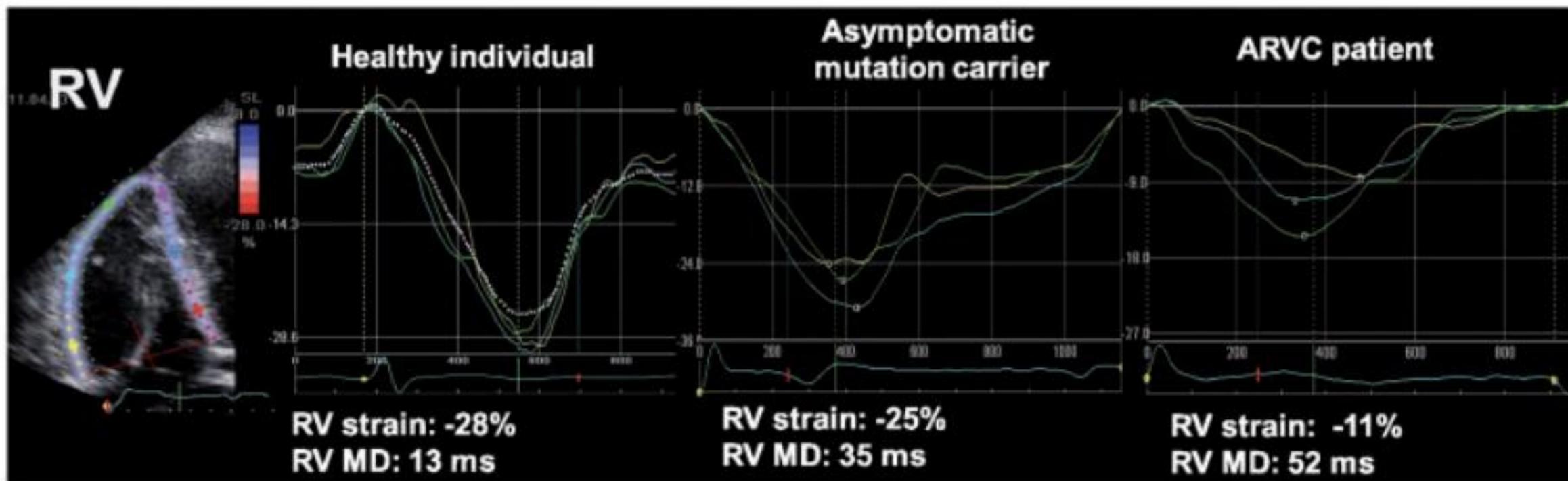


Strain

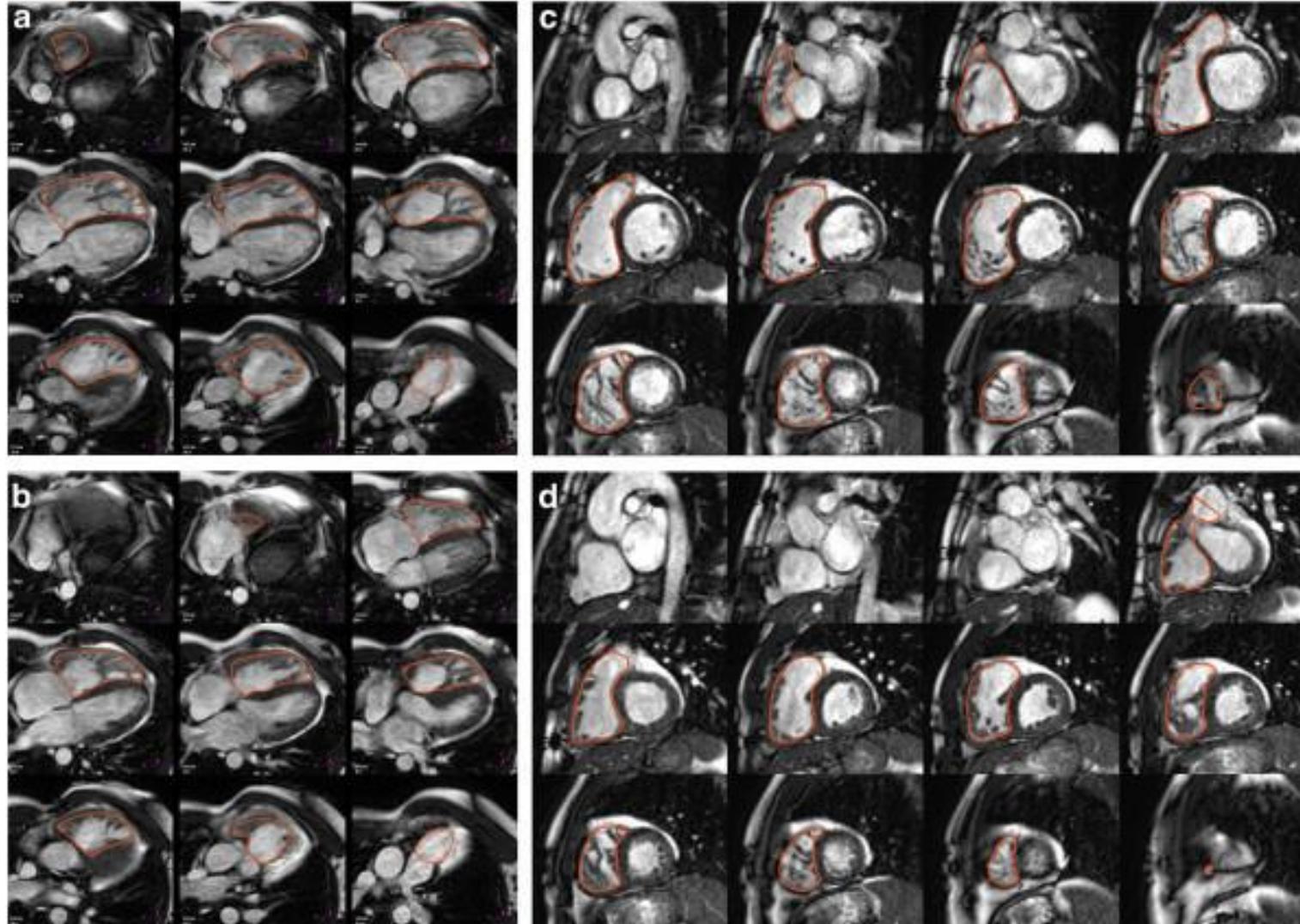


Comprehensive multi-modality imaging approach in arrhythmogenic cardiomyopathy— an expert consensus document of the European Association of Cardiovascular Imaging

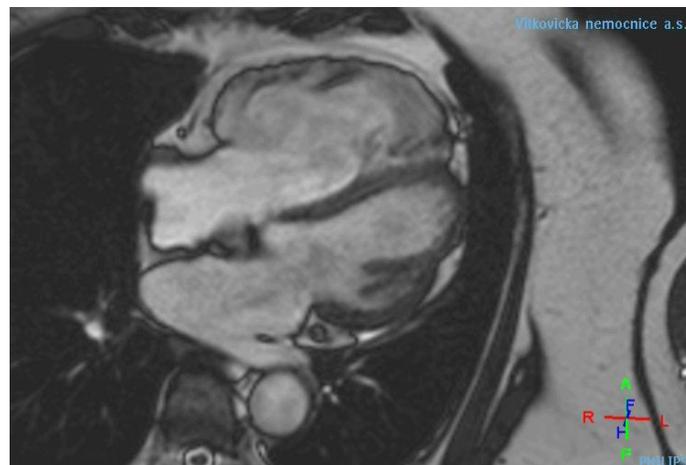
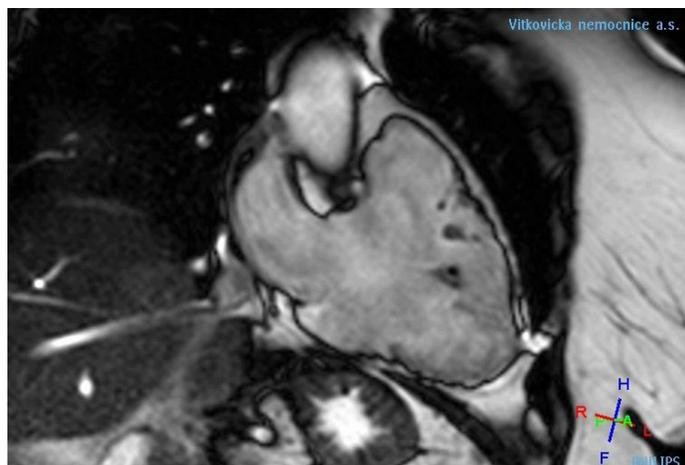
Kristina H. Haugaa^{1*}, Cristina Basso², Luigi P. Badano³, Chiara Bucciarelli-Ducci⁴,
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Juhani Knuuti⁹, Patrizio Lancellotti¹⁰, William McKenna¹¹, Danilo Neglia¹²,
Bogdan A. Popescu¹³, Thor Edvardsen¹



CMR



CMR



Ventricular dilatation and systolic dysfunction by CMR (nomograms for age, sex, and BSA).

Right ventricular dilatation and systolic dysfunction

	Women	Men	Athletes
EDV/ BSA (ml/m ²)	>112	>121	>130
EF (%)	<51	<52	<52

Right Ventricular Volume

Age: years Sex:

Height: cm Weight: kg BSA: m²

EDV: ml ESV: ml Mass: g

EF: **46%** (50-76)

EDV	ESV	SV	Mass
292 ml	159 ml	133 ml	0 g
(121-221)	(34-94)	(74-142)	(40-97)

EDVI	ESVI	SVI	Mass index
141 ml/m ²	77 ml/m ²	64 ml/m ²	0 g/m ²
(65-111)	(18-47)	(39-71)	(22-49)

Multimodality Imaging in Arrhythmogenic Right Ventricular Cardiomyopathy

Nitin Malik, MD; Monica Mukherjee, MD, MPH; Katherine C. Wu, MD; Stefan L. Zimmerman, MD; Junzhen Zhan, MD; Hugh Calkins, MD; Cynthia A. James, PhD; Nisha A. Gilotra, MD; Farooq H. Sheikh, MD; Harikrishna Tandri, MD; Shelby Kutty, MD, PhD; Allison G. Hays, MD

Feature tracking

RSNA

Right Ventricular Strain Derived from Cardiac MRI Feature Tracking for the Diagnosis and Prognosis of Arrhythmogenic Right Ventricular Cardiomyopathy

Key Result

Right ventricular global longitudinal strain (RV GLS) derived from cardiac MRI feature tracking demonstrated good diagnostic and prognostic value in arrhythmogenic right ventricular cardiomyopathy (ARVC).

Patients:

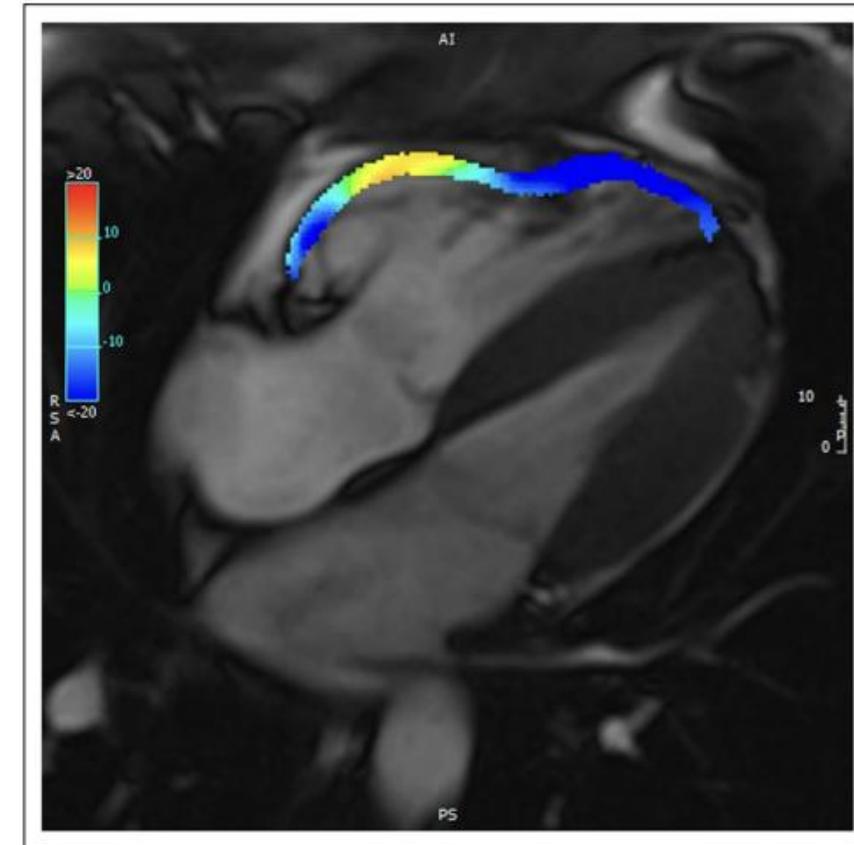
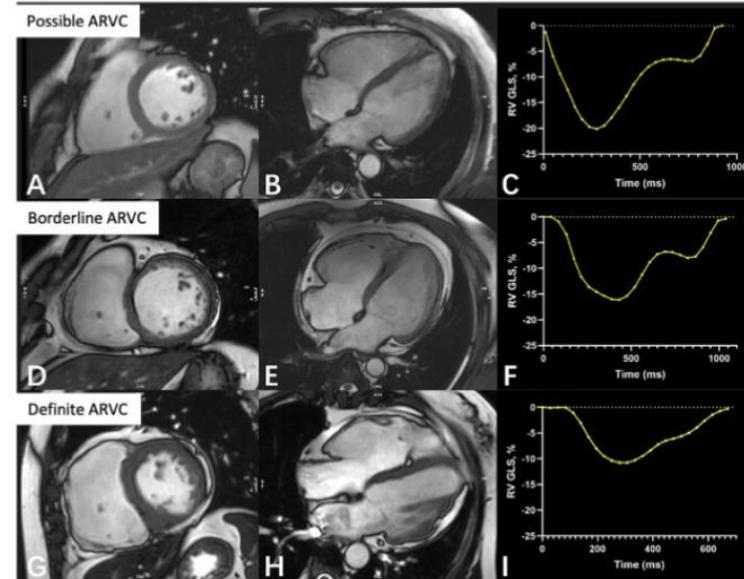
- 247 patients diagnosed with ARVC based on the revised Task Force criteria (possible: $n = 25$; borderline: $n = 40$; definite: $n = 182$)

Methods:

- Biventricular global strain parameters calculated using cardiac MRI feature tracking were compared across ARVC diagnostic groups.
- Association of RV GLS with occurrence of cardiovascular events was assessed.

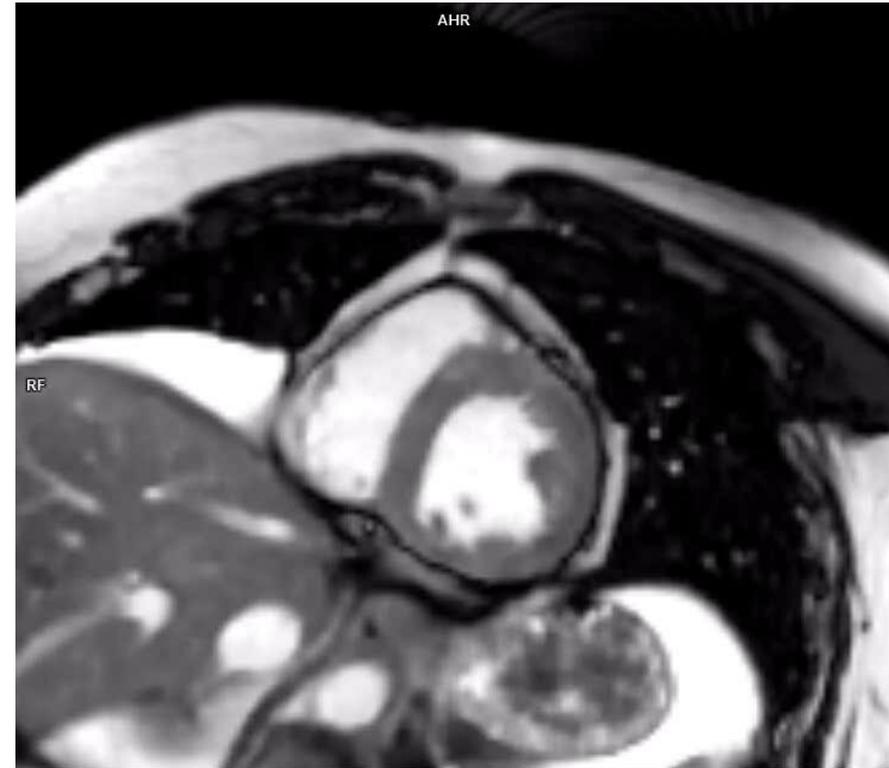
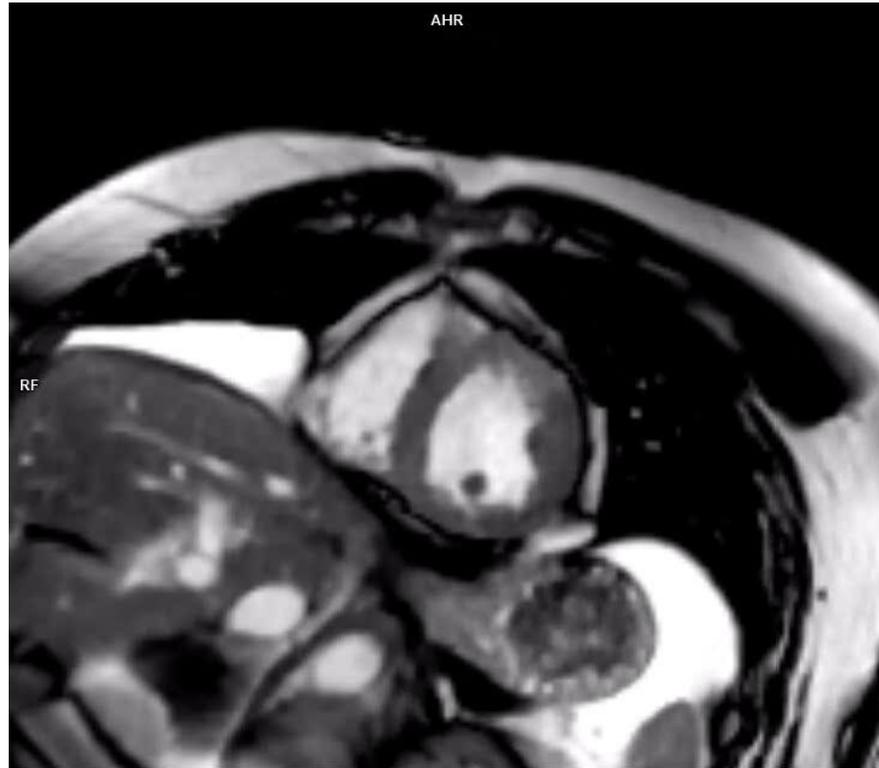
Results:

- Patients with definite ARVC had significantly reduced RV strain compared with possible or borderline groups.
- RV GLS was independently associated with occurrence of cardiovascular events (HR, 1.15; $P < .001$).



Porucha kinetiky a funkce PK

retrosternální srůsty



Strukturální změny

II Tissue characterisation of walls

MAJOR
Fibrofatty replacement of myocardium on endomyocardial biopsy

199
4

II. Tissue characterization of wall	
Major	
• Fibrofatty replacement of myocardium on endomyocardial biopsy	• 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	
	• Residual myocytes 60% to 75% by morphometric analysis (or 50% to 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

201
0

II. Structural myocardial abnormalities

By CE-CMR:

Major

- Transmural LGE (stria pattern) of ≥1 RV region(s) (inlet, outlet, and apex in 2 orthogonal views)

By EMB (limited indications):

Major

- Fibrous replacement of the myocardium in ≥1 sample, with or without fatty tissue

202
0

II. Structural alterations

Major

- Fibrous replacement of the myocardium in ≥1 sample, with or without fatty tissue, at histology

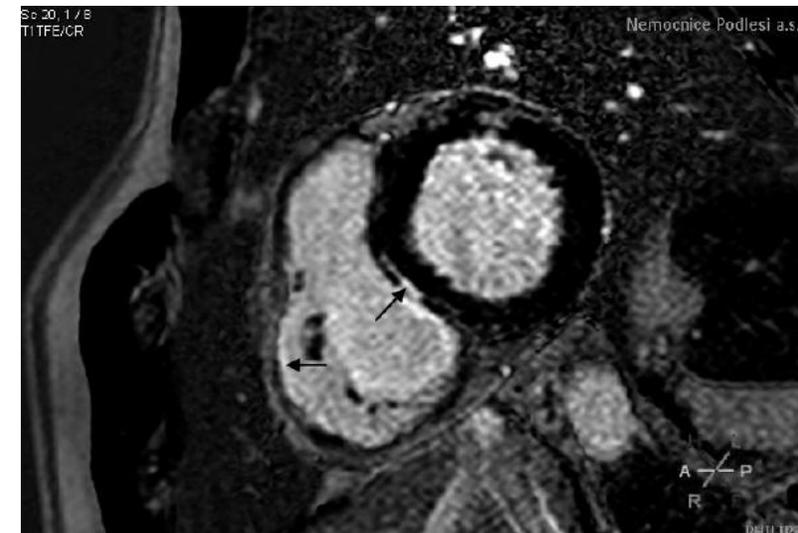
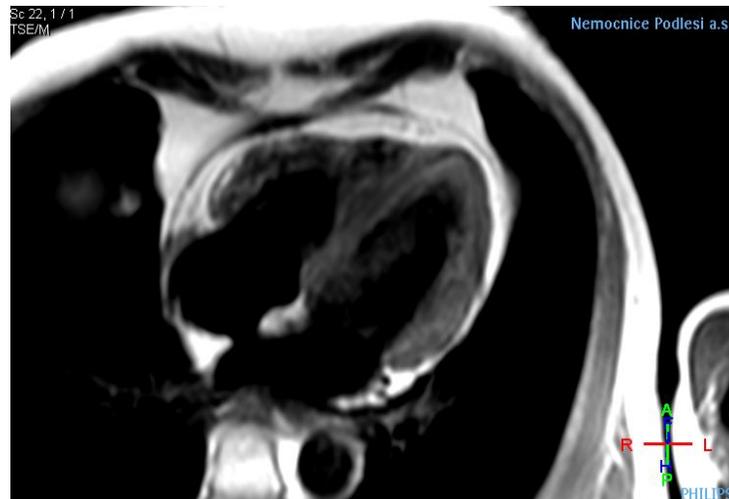
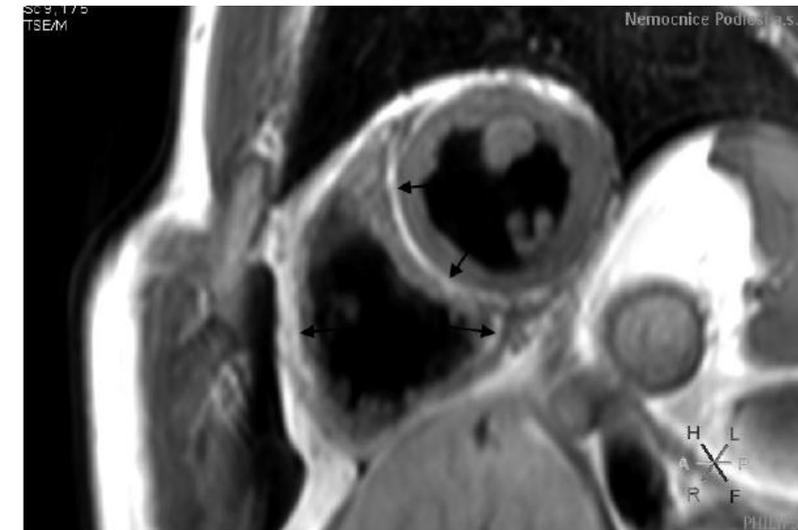
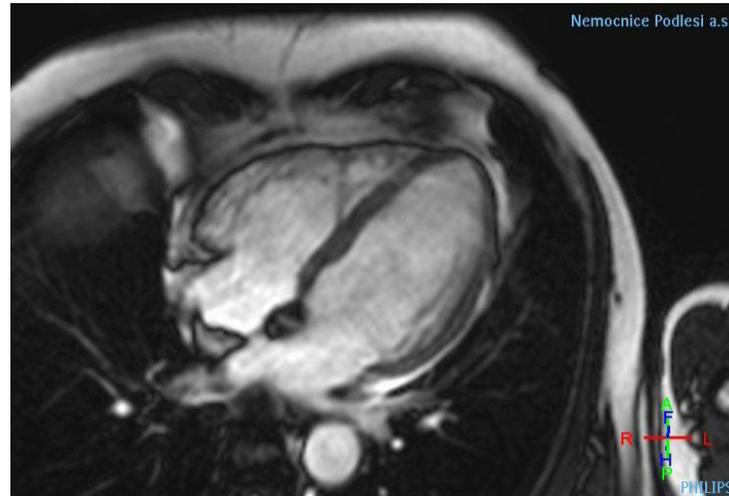
Minor

- Unequivocal RV LGE (confirmed in 2 orthogonal views) in ≥1 RV region(s) (excluding tricuspid valve)

202
4

Tukově-fibrózní náhrada myokardu

- kombinace poruchy kinetiky s odpovídající tukově-fibrózní náhradou – až 98 % přesnost v diagnostice ARVC



The
American Journal
of Cardiology

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CARDIOMYOPATHY - Volume 118, Issue 11, P1730-1736, December 01, 2016

Usefulness of Combined Functional Assessment by Cardiac Magnetic Resonance and Tissue Characterization Versus Task Force Criteria for Diagnosis of Arrhythmogenic Right Ventricular Cardiomyopathy

Giovanni Donato Aquaro, MD, PhD, Andrea Barison, MD, Giancarlo Todiere, MD, ... - Gianluca Di Bella, MD, PhD, Michele Emdin, MD, PhD, Pierluigi Festa, MD, ... Show more

Affiliations & Notes Article Info

CMR

te Riele et al. *Journal of Cardiovascular Magnetic Resonance* 2014, 16:50
<http://jcmr-online.com/content/16/1/50>

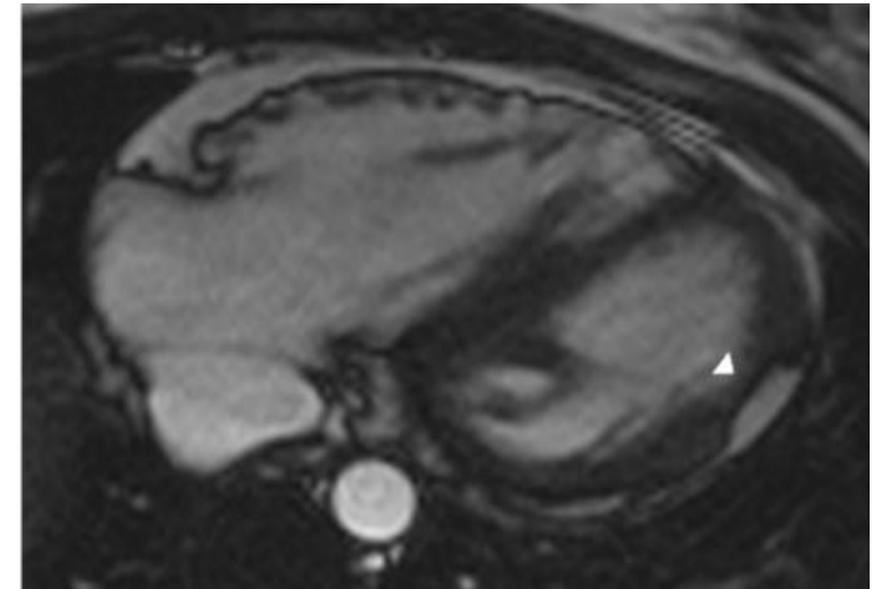
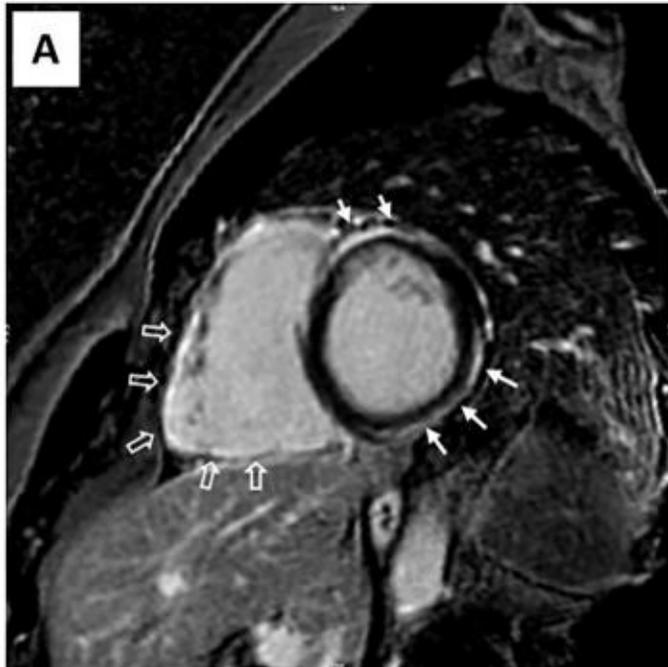
 **Journal of Cardiovascular
Magnetic Resonance**

REVIEW

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Arrhythmogenic right ventricular cardiomyopathy (ARVC): cardiovascular magnetic resonance update

Anneline SJM te Riele^{1,2}, Harikrishna Tandri² and David A Bluemke^{3,4*}



Biv-ACM

„like“ pattern

„bite-

Doporučení pro... | Guidelines

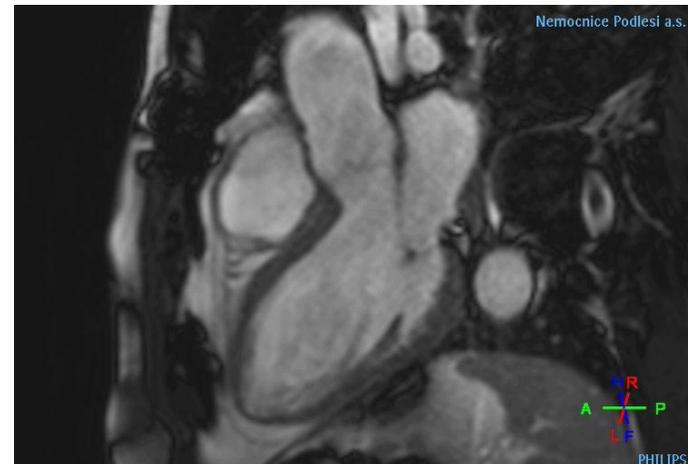
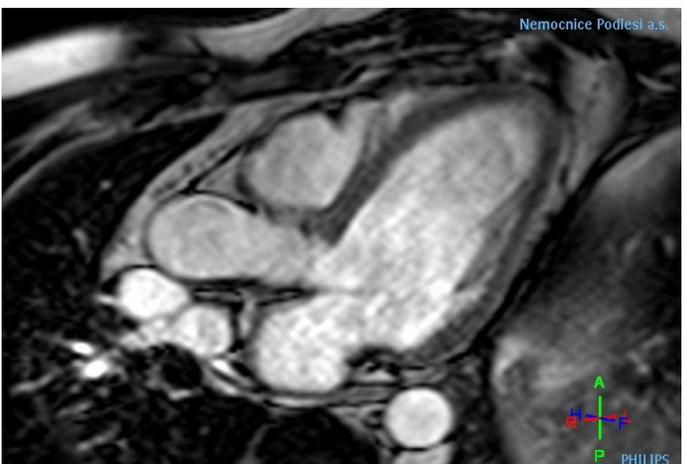
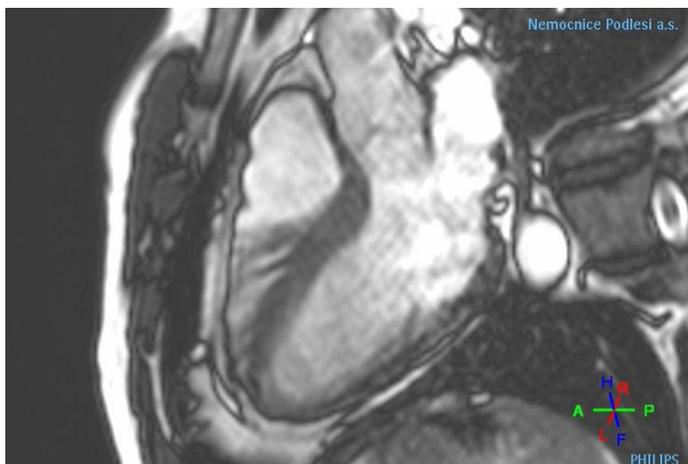
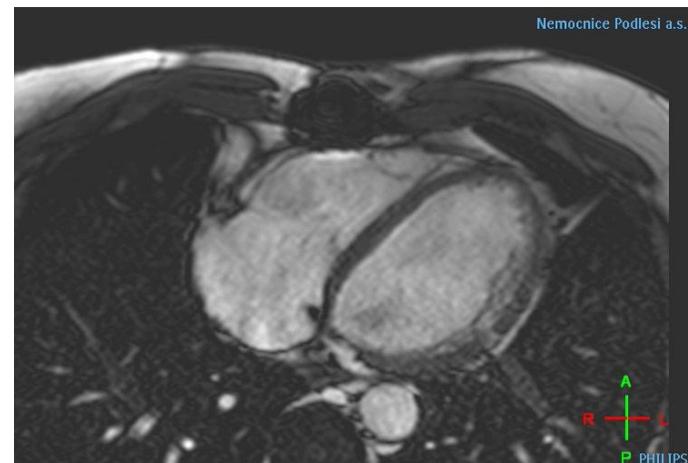
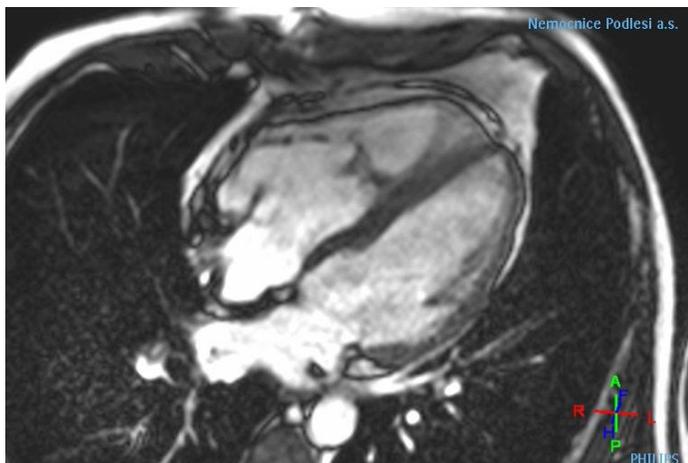
Doporučení ESC pro diagnostiku a léčbu pacientů s komorovými arytmiemi a prevenci náhlé srdeční smrti 2022.

Zeppenfeld K, Tfelt-Hansen J, de Riva M, Winkel BG, Behr ER, Blom NA, Charron P, Corrado D, Dagres N, de Chillou C, Eckardt L, Friede T, Haugaa KH, Hocini M, Lambiase PD, Marijon E, Merino JL, Peichl P, Priori SG, Reichlin T, Schulz-Menger J, Sticherling C, Tzeis S, Verstrael A, Volterrani M; ESC Scientific Document Group.

Překlad zkráceného dokumentu připravený Českou kardiologickou společností

Tabulka 29 doporučení – Doporučení pro diagnostiku, rizikovou stratifikaci, prevenci náhlé srdeční smrti a léčbu komorových arytmií u pacientů s arytmogenní kardiomyopatií pravé komory

Doporučení	Třída ^a	Úroveň ^b
Diagnostické zhodnocení a obecná doporučení		
U pacientů s podezřením na ARVC je doporučeno provedení CMR.	I	B
U pacientů s podezřením na ARVC nebo u pacientů s definitivní diagnózou ARVC je doporučeno genetické poradenství a testování.	I	B
U pacientů s definitivní diagnózou ARVC je doporučen zákaz cvičení s vysokou intenzitou.	I	B
Zákaz cvičení s vysokou intenzitou ^c může být zvážen u nositelů patogenních mutací pro ARVC bez fenotypického projevu.	IIb	C
Léčba betablokátry může být zvážena u všech pacientů s definitivní diagnózou ARVC.	IIb	C

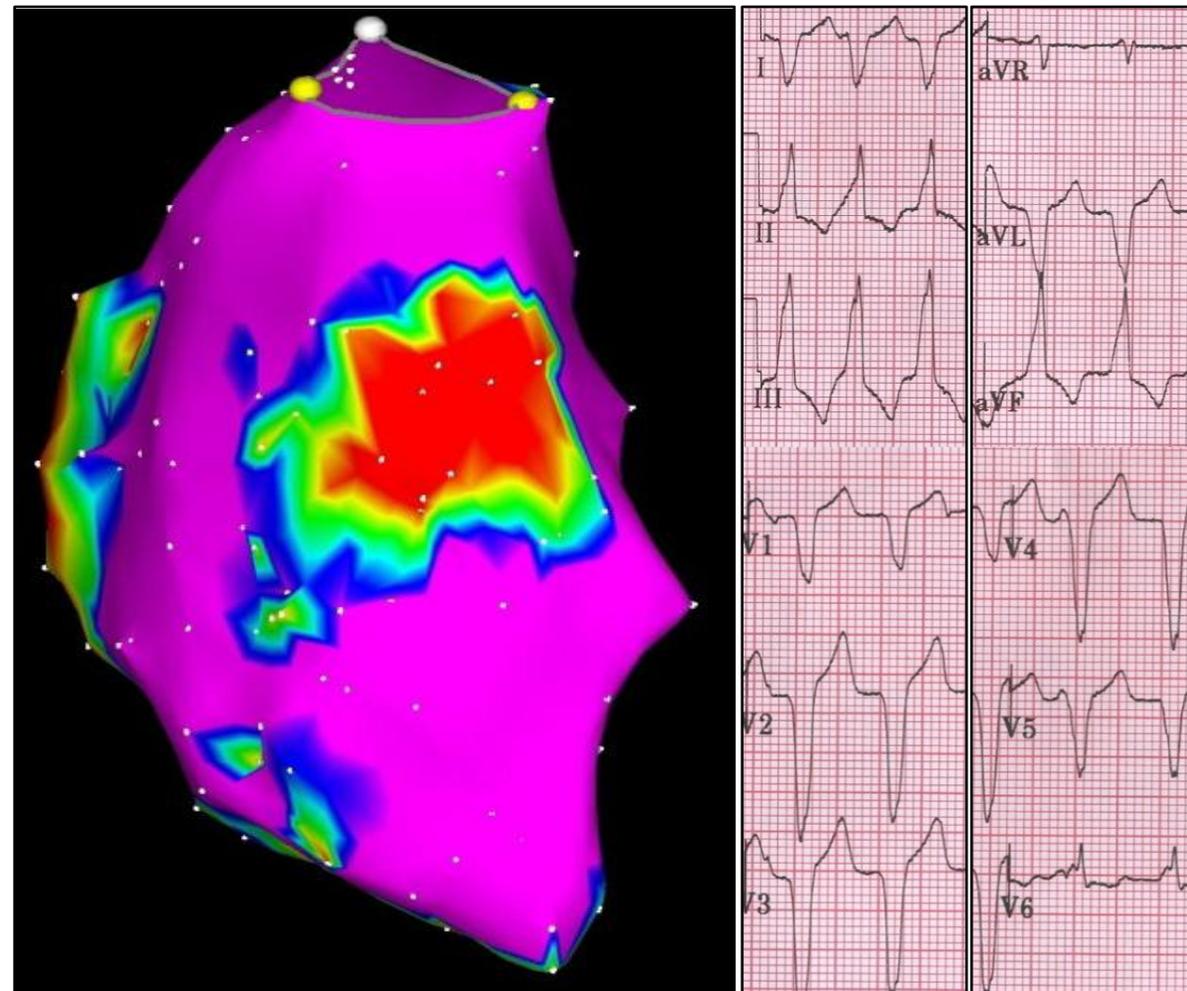
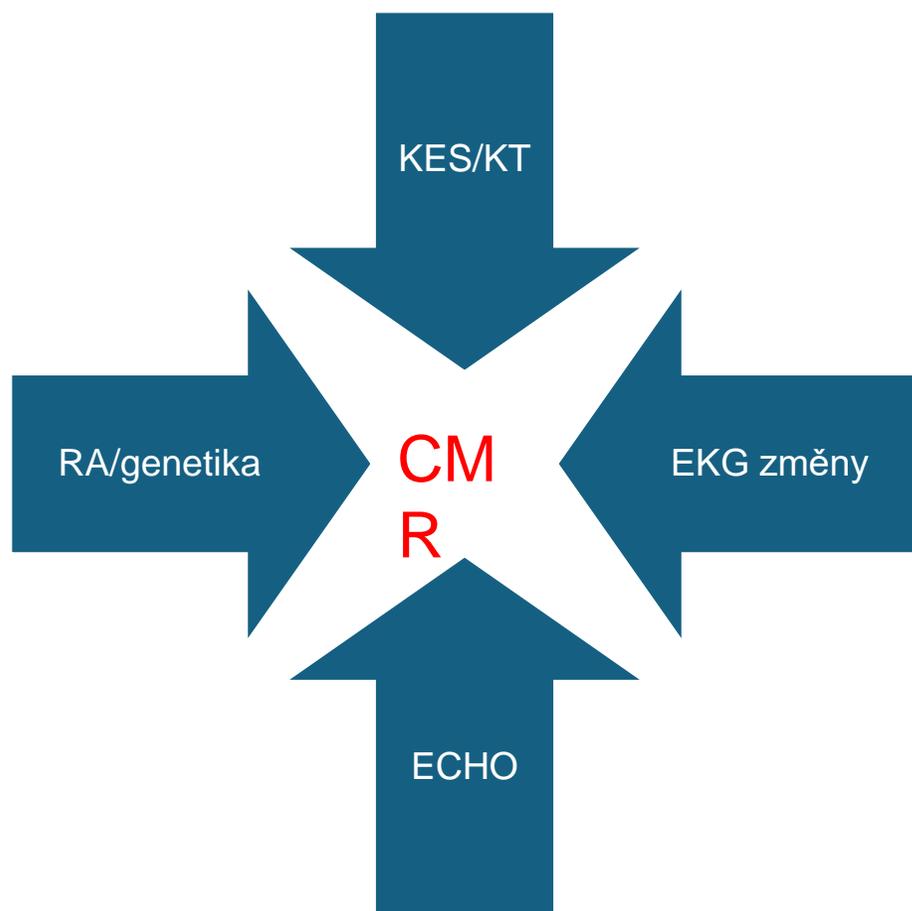


potvrzená ARVC (r. 2012)
(r.2019) - mutace PKP2

bratr (r.2017)

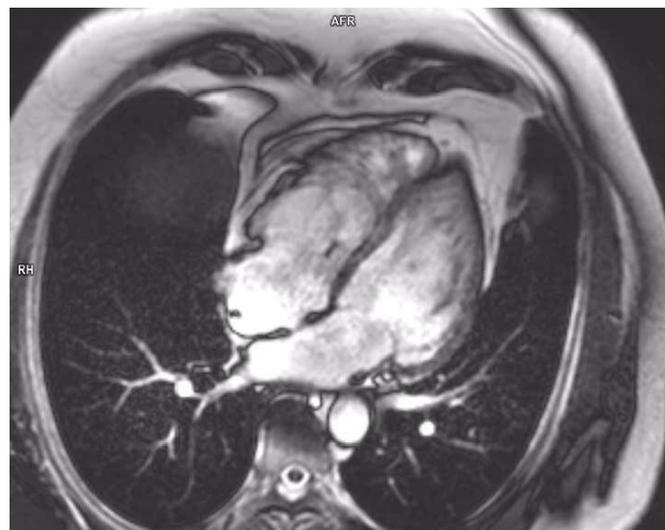
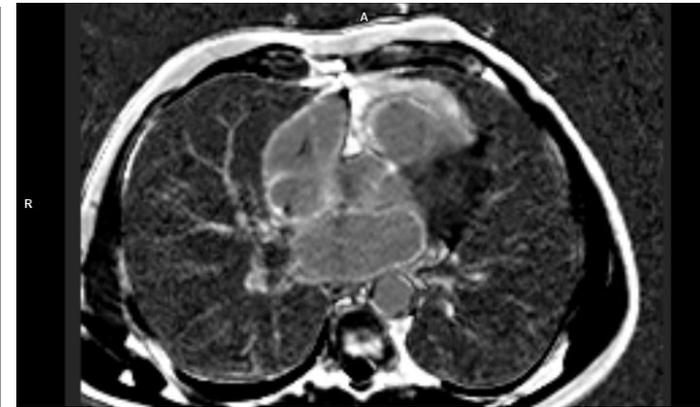
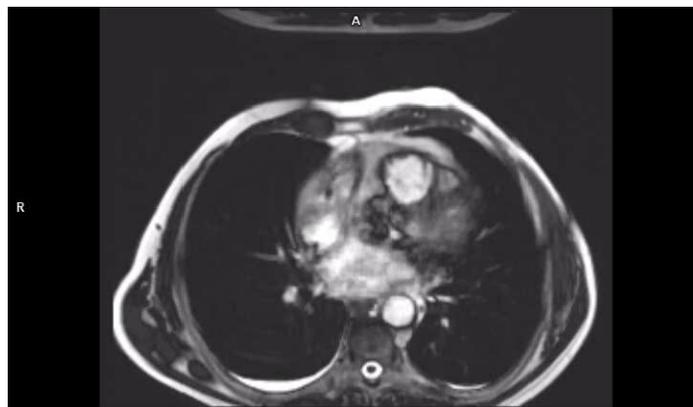
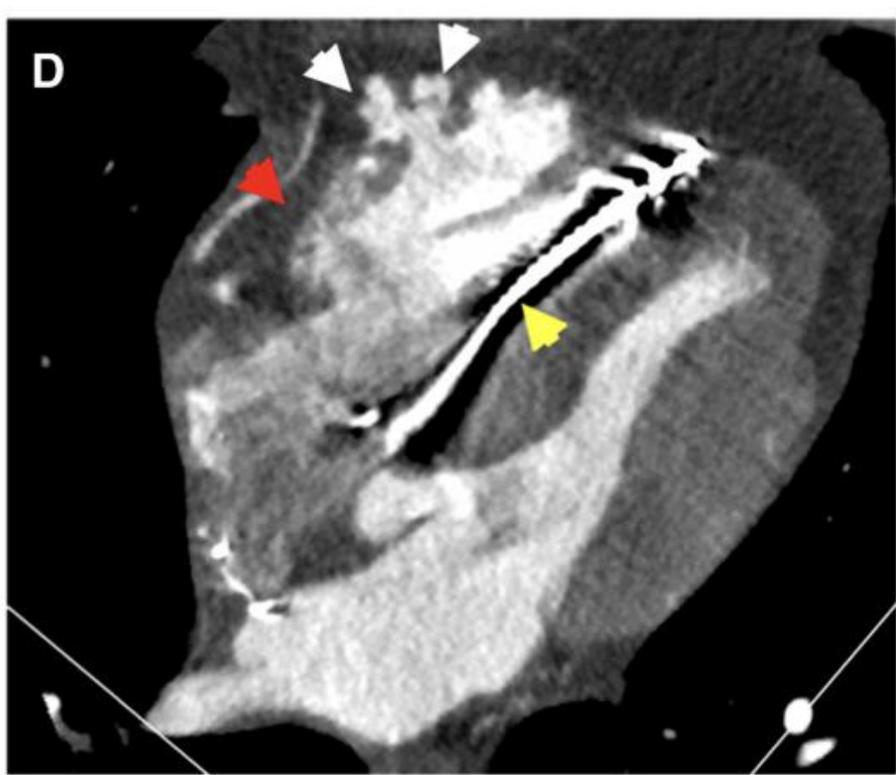
syn bratra

Diagnostický postup



CT

- při kontraindikaci CMR

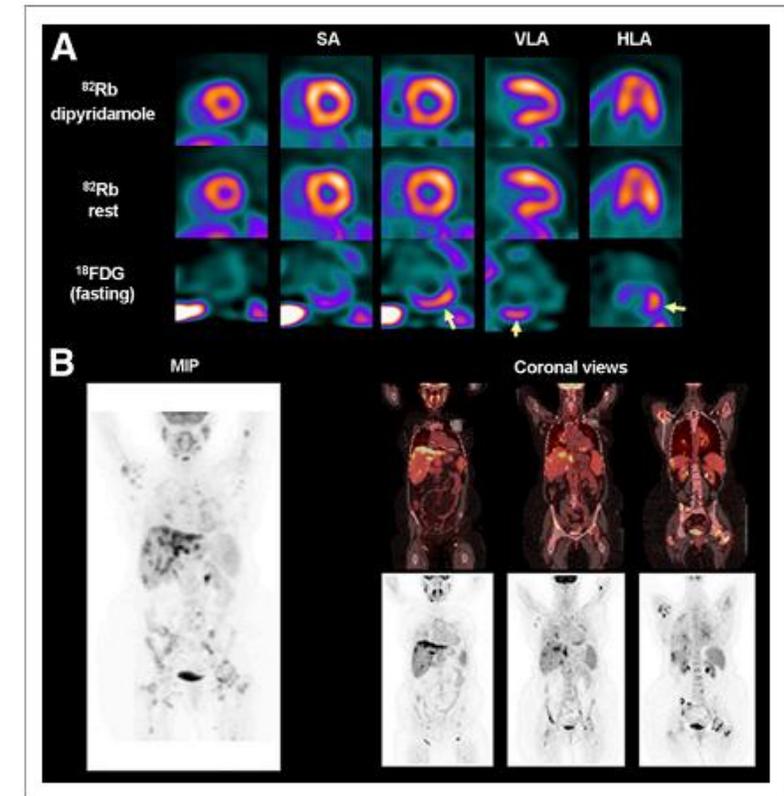


ADVANCES IN CARDIOVASCULAR IMAGING

Multimodality Imaging in Arrhythmogenic Right Ventricular Cardiomyopathy

Nitin Malik, MD; Monica Mukherjee, MD, MPH; Katherine C. Wu, MD; Stefan L. Zimmerman, MD; Junzhen Zhan, MD; Hugh Calkins, MD; Cynthia A. James, PhD; Nisha A. Gilotra, MD; Farooq H. Sheikh, MD; Harikrishna Tandri, MD; Shelby Kutty, MD, PhD; Allison G. Hays, MD

Overlap diagnosis	Imaging modality			
	Echocardiography	CMR	MDCT	PET
Athlete's heart	Balanced dilation of cardiac chambers	Same structural findings as echocardiography LGE, if present, is generally limited to the RV insertion points
	Wall thickness <15 mm			
	Absence of RWMA			
Sarcoidosis	Reduced LV function	Characteristic nonischemic LGE pattern of the interventricular septum and/or lateral wall, often basal	Mediastinal lymphadenopathy may be detected	Heterogenous tracer uptake in active cardiac sarcoid
	RWMA may be seen			
Myocarditis	Reduced LV function	Reduced LV function	...	May demonstrate and localize active myocardial inflammation
		RWMA may be seen		
		Tissue edema		
	RWMA may be seen	Patchy subepicardial myocardial LGE (nonischemic pattern)		
		May show pericardial involvement		
Dilated cardiomyopathy	Dilated LV cavity	Dilated LV cavity
		Reduced LV function		
	Reduced LV function	Mid-wall septal LGE on CMR		
RVOT-VT	Normal imaging findings	Normal imaging findings
Brugada syndrome	Normal imaging findings	Normal imaging findings
Uhl's anomaly (total absence of right ventricular myocardium)	Apposition of endocardium and epicardium	Apposition of endocardium and epicardium



Advanced Imaging of Cardiac Sarcoidosis

Imke Schatka and Frank M. Bengel

Department of Nuclear Medicine, Hannover Medical School, Hannover, Germany

CMR a prognóza

ARVC Risk Calculator v3.0

The Arrhythmogenic Right Ventricular Cardiomyopathies (ARVC) Risk Calculator estimates the risk of ventricular arrhythmias (VA) within 5 years for patients with a *definite* ARVC diagnosis. Please read all definitions below carefully, as well as our general [disclaimer](#), to avoid invalid interpretation of results.

This is

Prim: Right ventricular ejection fraction (%)

– Ris

– Ris

50

All pa: *As measured by cardiac MRI.*

– Risk of fast sustained VA (only ventricular tachycardia (VT) <200bpm, ventricular fibrillation/flutter (VF), sudden cardiac arrest/death (SCA/SCD))

CMR a prognóza

150 pts

- 41 % – ARVC
- 37 % – ABVC
- 12 % – ALVC
- 10 % – negat. CMR

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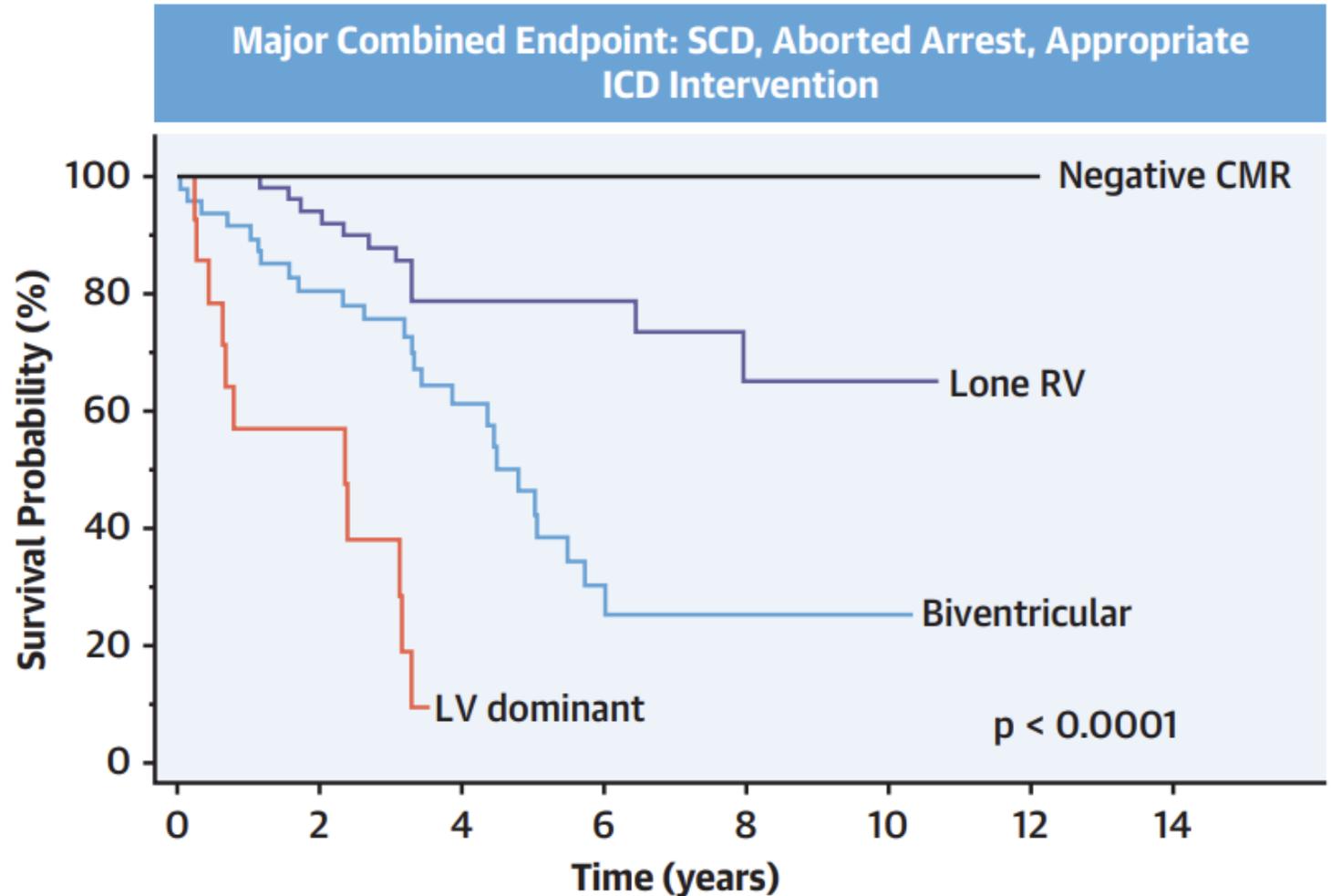
VOL. 75, NO. 22, 2020

ORIGINAL INVESTIGATIONS

Prognostic Value of Magnetic Resonance Phenotype in Patients With Arrhythmogenic Right Ventricular Cardiomyopathy



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Francesco Bianco, MD,² Nicoletta Botto, BSc,³ Pierluigi Lesizza, MD,³ Crysanthos Grigoratos, MD, PhD,⁴
Monia Minati, MD,⁵ Matteo Dell'Omodarme, PhD,⁶ Alessandro Pingitore, MD, PhD,⁶ Davide Stolfo, MD,⁵
Matteo Dal Ferro, MD,⁷ Marco Merlo, MD,¹ Gianluca Di Bella, MD, PhD,¹ Gianfranco Sinagra, MD¹



Stratifikace rizika

- 369 pts se suspekci na ARVC

Abnormální CMR:

- dilatace PK
- systolická dysfunkce PK
- porucha kinetiky
- tuková infiltrace
- LGE

Prognostic value of cardiovascular magnetic resonance in patients with suspected arrhythmogenic right ventricular cardiomyopathy

Monica Deac¹, Francisco Alpendurada, Fariba Fanaie, Raj Vimal, John-Paul Carpenter, Adelle Dawson, Chris Miller, Isabelle Roussin, Elisa di Pietro, Tefik F Ismail, Michael Roughton, Joyce Wong, Dana Dawson, Janice A Till, Mary N Sheppard, Raad H Mohiaddin, Philip J Kilner, Dudley J Pennell, Sanjay K Prasad

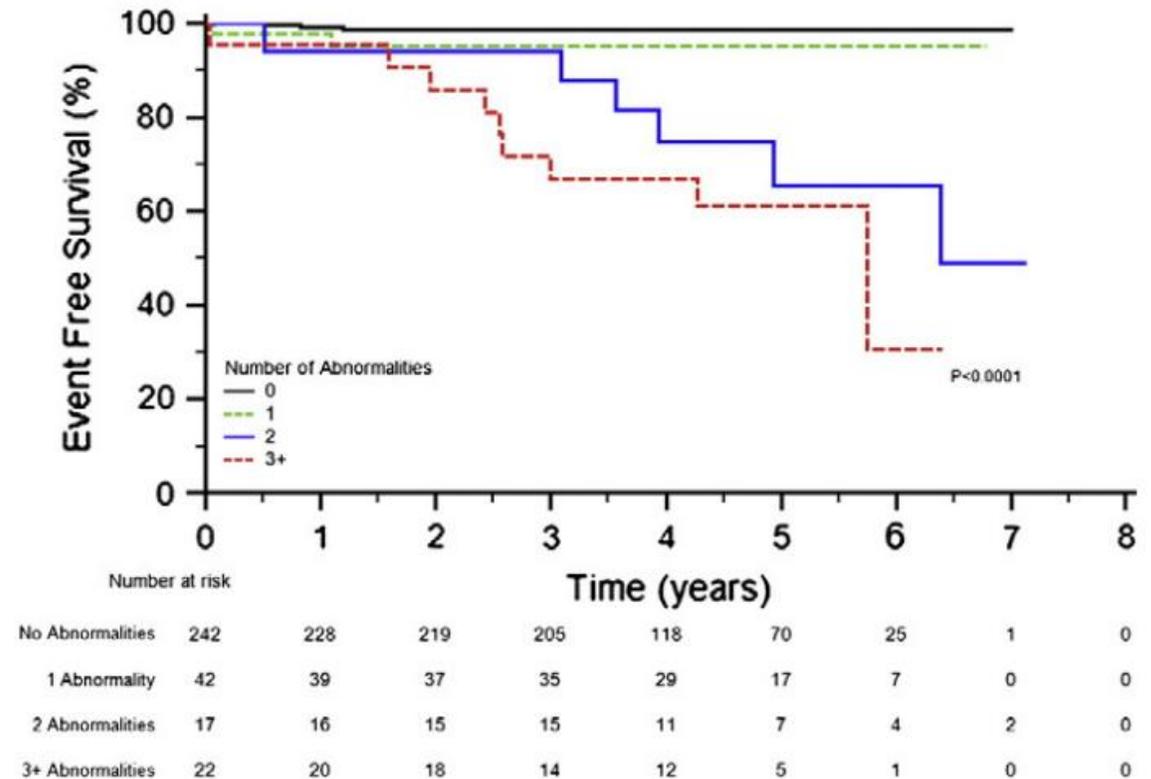


Fig. 6. Kaplan–Meier curves on event-free survival according to number of abnormalities found on CMR.



AGEL

Nemocnice AGEL
Trinec-Podlesí

Děkuji za pozornost

Nemocnice AGEL
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