800 ANNI 1222 • 2022

UNIVERSITÀ DEGLI STUDI DI PADOVA

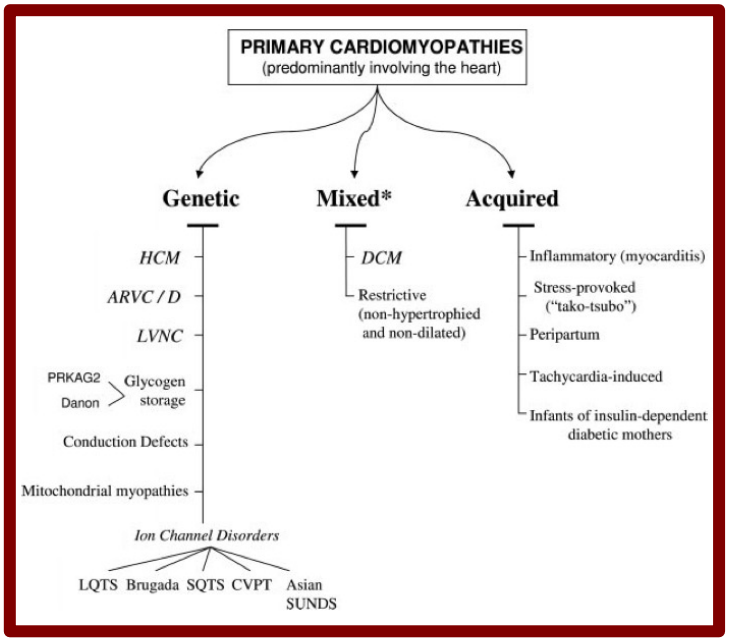
Cardiomiopatia Aritmogena

MASTER

Tecniche di acquisizione di risonanza magnetica cardiaca

AA 25_26

Classificazione



Secondary Cardiomyopathies

Infiltrative*
- Amyloidosis (primary, familial autosomal dominant†, senile, secondary forms)
- Gaucher disease†
- Hurler's disease†
- Hunter's disease†

Storage‡
- Hemochromatosis
- Fabry's disease†
- Glycogen storage disease† (type II, Pompe)
- Niemann-Pick disease†

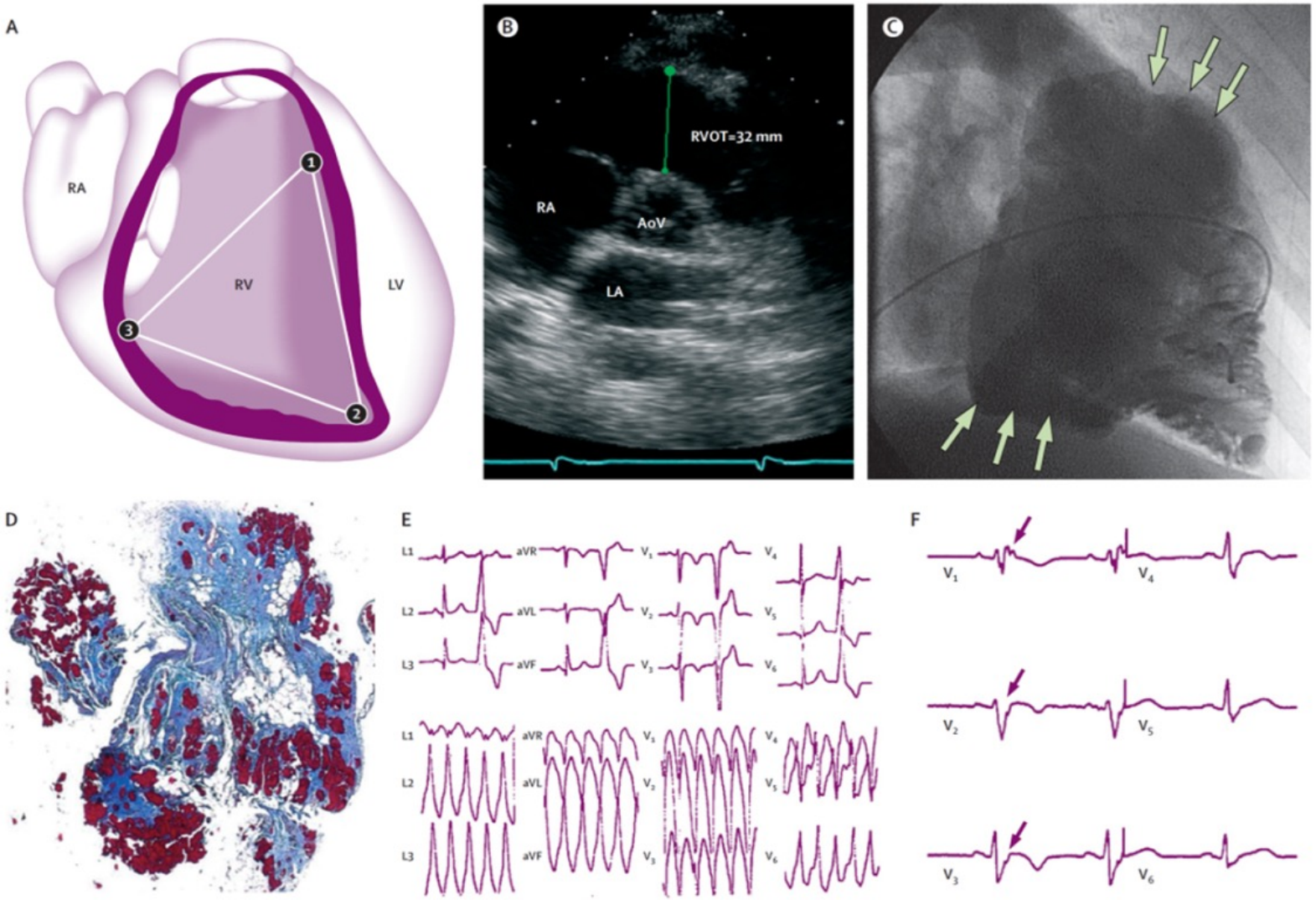
Toxicity
- Drugs, heavy metals, chemical agents

Endomyocardial
- Endomyocardial fibrosis
- Hypereosinophilic syndrome (Löeffler's endocarditis)

Inflammatory (granulomatous)
- Sarcoidosis

Circulation. 2006;113:1807-1816

Arrhythmogenic RV Cardiomyopathy

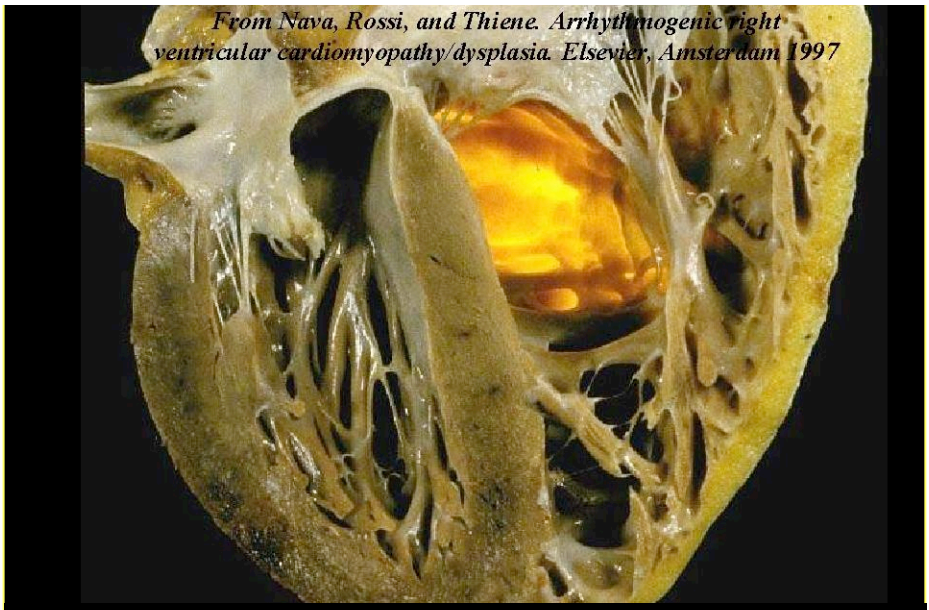


Basso C, Lancet 2009

Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia

Proposed Modification of the Task Force Criteria

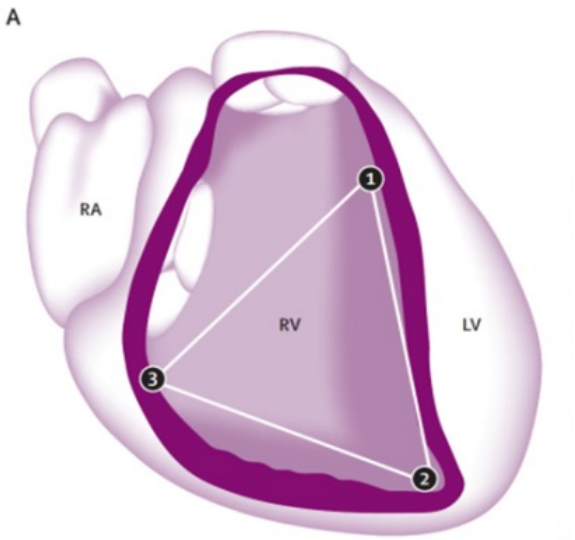
I. Global or regional dysfunction & structural alterations
II. Tissue characterization of wall
III. Repolarization abnormalities
IV. Depolarization/conduction abnormalities
V. Arrhythmias
VI. Family history



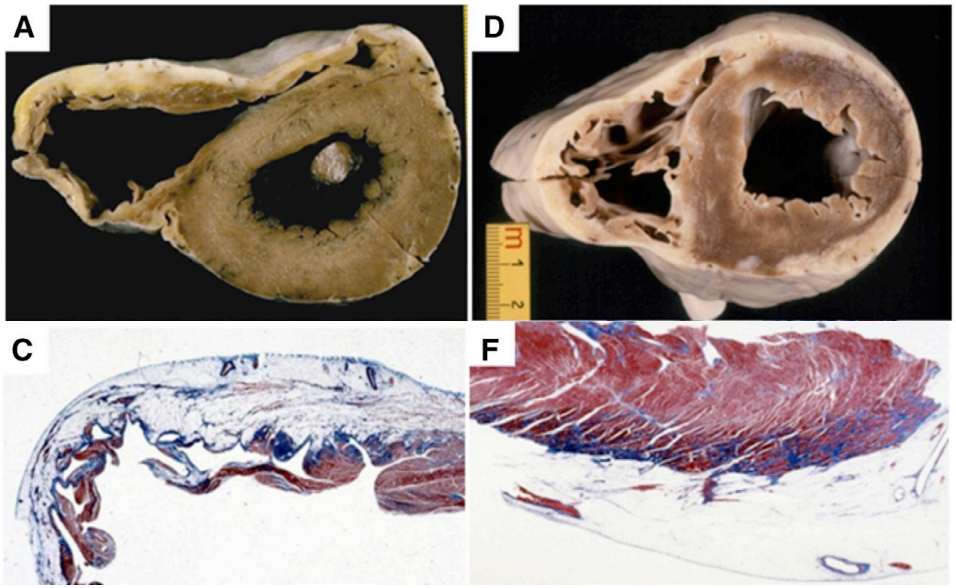
RV goals-oriented!!!

Definite/Borderline/Possible Diagnosis

From RV to LV



«**Triangle** of dysplasia»



«**Quadrangle** of ARVC» which also includes the LV infero-lateral wall

Corrado D. Circulation Research. 2017 (784–802)

L'imaging nei TF 1994

- Familial history of ARVC and/or sudden death
- ECG depolarization/conduction/repolarization abnormalities
- Arrhythmias of right ventricular origin
- Global and/or regional dysfunction and structural alterations of the right ventricle
- Fibro-fatty replacement of the right ventricular myocardium

McKenna et al, Br Heart J 1994

L'imaging nei Revised TF 2010

EUROPEAN SOCIETY OF CARDIOLOGY®
European Heart Journal (2010) **31**, 806–814
doi:10.1093/eurheartj/ehq025

SPECIAL REPORT

Diagnosis of arrhythmogenic right ventricular cardiomyopathy/dysplasia

Proposed Modification of the Task Force Criteria

Revision of the diagnostic criteria provides guidance on the role of emerging diagnostic modalities and advances in the genetics of ARVC/D. The criteria have been modified to incorporate new knowledge and technology to improve diagnostic sensitivity, but with the important requisite of maintaining diagnostic specificity. The approach of classifying structural, histological, electrocardiographic, arrhythmic, and genetic features of the disease as major and minor criteria has been maintained. In this modification of the Task Force criteria, quantitative criteria are proposed and abnormalities are defined on the basis of comparison with normal subject data.

Global or regional dysfunction and structural alterations

Major

By 2D echo:

- Regional RV akinesia, dyskinesia, or aneurysm
- *and* 1 of the following (end diastole):
  - PLAX RVOT ≥32 mm (corrected for body size [PLAX/BSA] ≥19 mm/m^2)
  - PSAX RVOT ≥36 mm (corrected for body size [PSAX/BSA] ≥21 mm/m^2)
  - *or* fractional area change ≤33%

By MRI:

- Regional RV akinesia or dyskinesia or dyssynchronous RV contraction
- *and* 1 of the following:
  - Ratio of RV end-diastolic volume to BSA ≥110 mL/m^2 (male) or ≥100 mL/m^2 (female)
  - *or* RV ejection fraction ≤40%

By RV angiography:

- Regional RV akinesia, dyskinesia, or aneurysm

Global or regional dysfunction and structural alterations

Minor

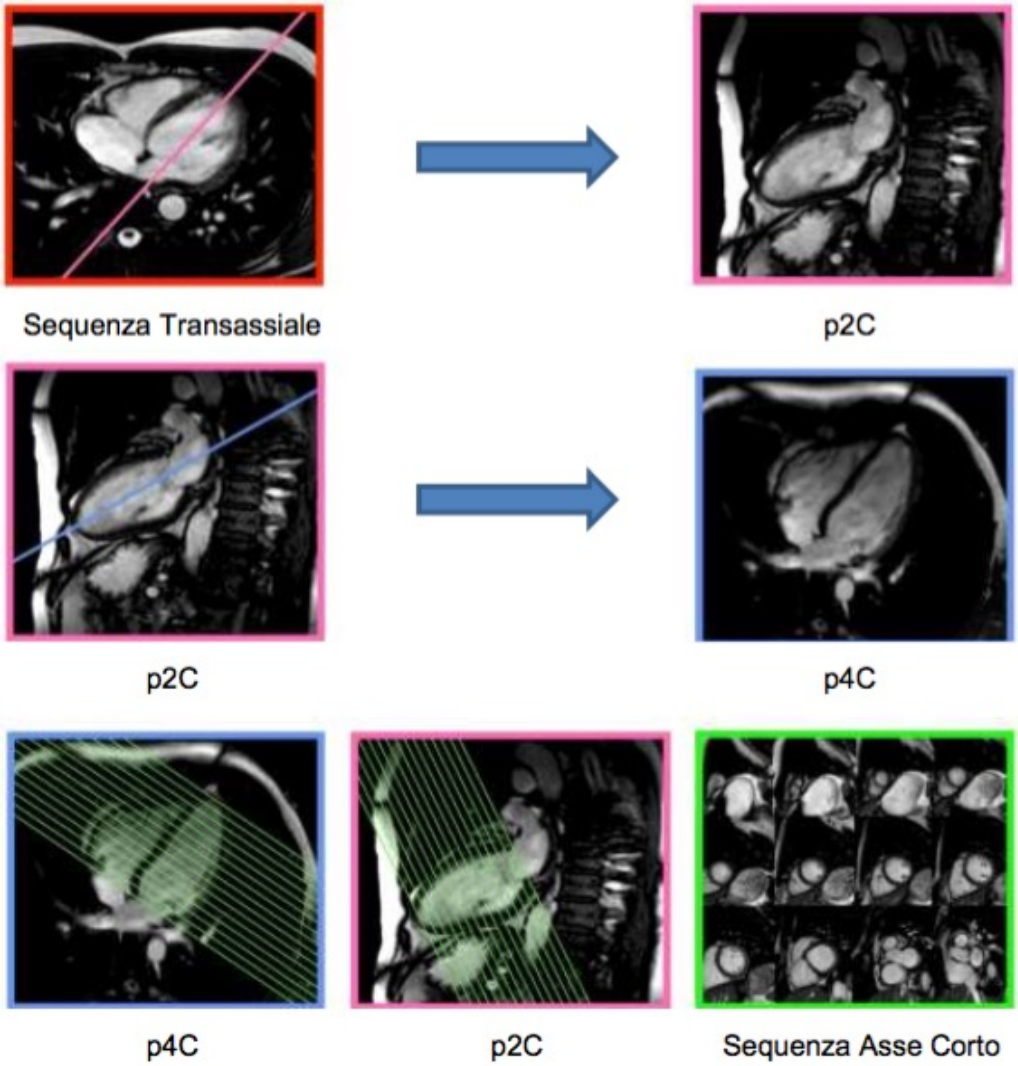
By 2D echo:

- Regional RV akinesia or dyskinesia
- *and* 1 of the following (end diastole):
  - PLAX RVOT ≥29 to <32 mm (corrected for body size [PLAX/BSA] ≥16 to <19 mm/m^2)
  - PSAX RVOT ≥32 to <36 mm (corrected for body size [PSAX/BSA] ≥18 to <21 mm/m^2)
  - *or* fractional area change >33% to ≤40%

By MRI:

- Regional RV akinesia or dyskinesia or dyssynchronous RV contraction
- *and* 1 of the following:
  - Ratio of RV end-diastolic volume to BSA ≥100 to <110 mL/m^2 (male) or ≥90 to <100 mL/m^2 (female)
  - *or* RV ejection fraction >40% to ≤45%

Tips & Tricks



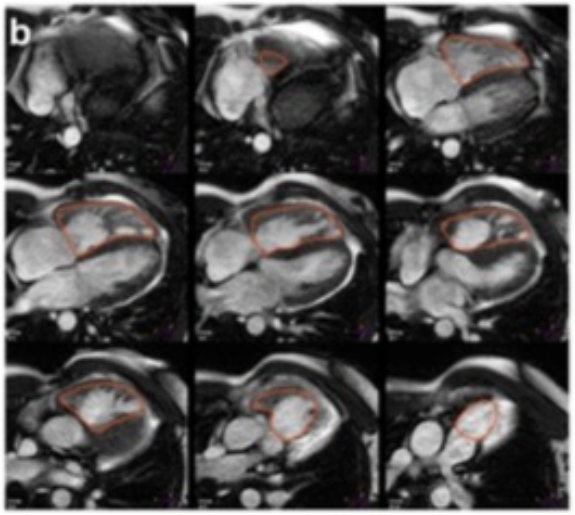
CMR pocket guide EACVI

Tips & Tricks

CMR pocket guide EACVI

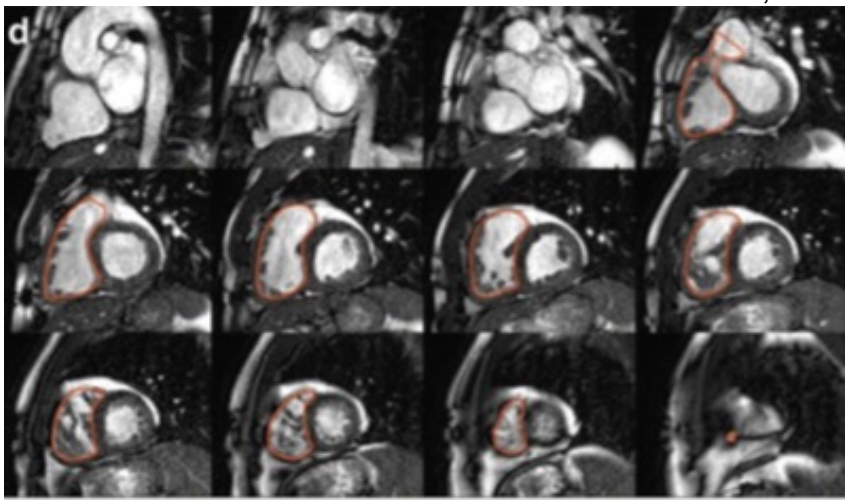
Funzione del Vdx

1. Sequenze pulsate **Cine SSFP** (imaging parallelo se necessario)
2. **Modulo cine trans-assiale** (dal diaframma alla biforcazione della polmonare) o **modulo cine Asse Corto** come per il VSin
   - Spessore fetta 6-8mm
   - Distanza tra le fette 0mm
3. Risoluzione temporale ≤ 45ms



Modulo trans-assiale

Schulz-Menger et al. Journal of Cardiovascular Magnetic Resonance 2013, 15:35



Modulo asse corto

Documenti di consenso

Visione dei cut-off

European Heart Journal - Cardiovascular Imaging (2019) 20, 1321–1331

Right ventricle ranges for adults aged 20–80 years

	Women				
	'Opposite'	Reference range	Mildly abnormal	Moderately abnormal	Severely abnormal
20–68 years					
EDV (mL)	<77	77–201	202–232	233–263	>263
EDV /BSA (mL/m^2)	<48	48–112	113–128	129–144	>144
ESV (mL)	<24	24–84	85–99	100–114	>114
ESV/BSA (mL/m^2)[b]	<12	12–52	53–62	63–72	>72
EF (%)	>71	51–71	41–51	30–40	<30
Mass (g)[b]	<21	21–49	50–56	57–63	>63
Mass/BSA (g/m^2)[b]	<12	12–28	29–32	33–36	>36

	Men				
	'Opposite'	Reference range	Mildly abnormal	Moderately abnormal	Severely abnormal
	<118	118–250	251–283	284–316	>316
	<61	61–121	122–136	137–151	>151
	<41	41–117	118–136	137–155	>155
	<19	19–59	60–69	70–79	>79
	>72	52–72	41–52	30–40	<30
	<25	25–57	58–65	66–73	>73
	<13	13–29	30–33	34–37	>37

Using the statistical method:

- **normal** range is defined as ±2 SD from the mean,
- **mildly** abnormal from this cut-off to 3 SD,
- **moderately** abnormal from the mild cut-off to 4 SD
- **severe** being more than 4 SD from the population mean.

- The term '**Opposite**' refers to values that are outside the normal range but in the opposite direction of typical thought of as pathology, e.g. smaller LV end-diastolic volumes or supra-normal LVEF.

Documenti di consenso

Visione dei cut-off

European Heart Journal - Cardiovascular Imaging (2019) 20, 1321–1331

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ESV (mL)	<24	24–84	85–99	100–114	>114	<41	41–117	118–136	137–155	>155
ESV/BSA (mL/m^2)[b]	<12	12–52	53–62	63–72	>72	<19	19–59	60–69	70–79	>79
EF (%)	>71	51–71	41–51	30–40	<30	>72	52–72	41–52	30–40	<30
Mass (g)[b]	<21	21–49	50–56	57–63	>63	<25	25–57	58–65	66–73	>73
Mass/BSA (g/m^2)[b]	<12	12–28	29–32	33–36	>36	<13	13–29	30–33	34–37	>37

Using the statis

- **normal** ran
- **mildly** abn
- **moderately**
- **severe** bei

- The term '**C** thought of a pathology, e.g. smaller LV end-diastolic volumes or supra-normal LVEF

By MRI:

- Regional RV akinesia or dyskinesia or dyssynchronous RV contraction
- *and* 1 of the following:
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  - or RV ejection fraction ≤40%

Documenti di consenso

Limitazioni e pregi

European Heart Journal - Cardiovascular Imaging (2019) 20, 1321–1331

Limiti

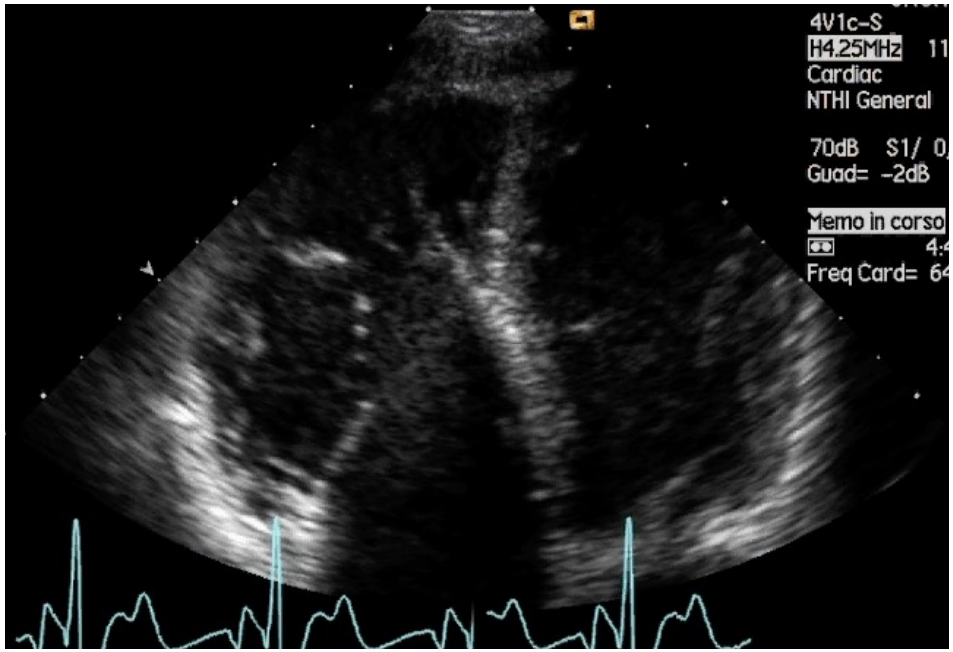
- Range molto ampio di età (20-80 aa);
- Esclusi i teenagers → problema ARVC;
- Non differenze in base all'etnia;
- Non presenti valori diversi per la popolazione degli atleti.

Pregi

- Più semplice il confronto con i valori di riferimento;
- Uniformità e semplificazione del post-processing;
- Il «grading» offre maggiore vicinanza alla pratica clinica.

Esempio:
FA 26%

Echo Limitations



- Acoustic window;
- Complexity of right ventricular geometry;
- Correct volumes/function evaluation;
- No information on tissue characterization

FA ≤ 33% (major)
FA 33-40 mm (minor)

CMR Advantages

- Evaluation of RV volumes/function
- Evaluation of RV wall and wall motion abnormalities
- Evaluation of Fatty Infiltration;
- Evaluation of Fibrosis

Protocollo RM

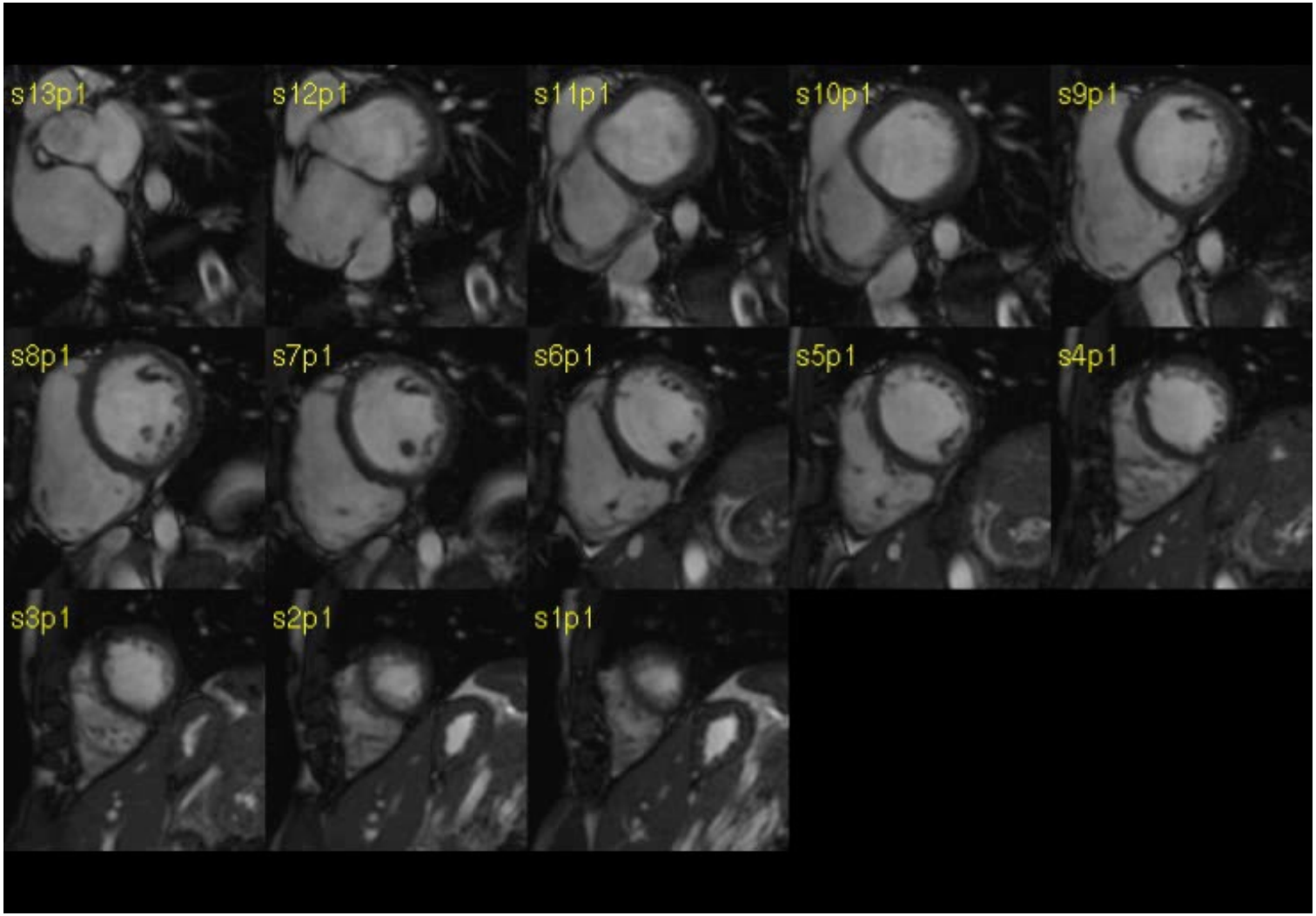
Sequence	Imaging plane	Parameters	Comments
Double inversion recovery TSE/FSE *a) Axial: with and without fat suppression* *b) short axis: without fat suppression*	a) Axial: obtain ~6-8 images centered on the left/right ventricle b) Short axis: obtain ~6-8 images centered on the left ventricle	TR = 2 R-R intervals, TE = 5 msec (minimum-full) (GE), TE = 30 msec (Siemens) slice thickness = 5 mm, interslice gap = 5 mm, and field of view (FOV) = 28–34 cm. ETL 16-24	This sequence provides optimal tissue characterization of the RV free wall. Prescribe from the pulmonary artery to the diaphragm. Fat suppression improves reader confidence in diagnosis of RV fat infiltration.
SSFP Bright Blood Cine Images	Axial, Four chamber and Short Axis. RV 3 chamber (optional)	TR/TE minimum, flip angle = 45-70°, slice thickness = 8 mm, interslice gap = 2 mm. FOV = 36–40 cm, 16–20 views per segment. Parallel imaging n = 2 is desirable	Axial images are best to assess RV wall motion. RV quantitative analysis is performed on the short axis cine images.
GADOLINIUM IS ADMINISTERED			**(usually 0.15 – 0.2 MMOL/KG)**
TI scout	Four chamber		TI scout sequences or trial TI times to suppress normal myocardium for the right inversion time.
Delayed Gadolinium Imaging (Phase Sensitive Inversion Recovery recommended)	Axial, Short Axis, Four Chamber and Vertical Long Axis	TR/TE per manufacturer recommendations flip angle = 20-25°, slice thickness = 8 mm, interslice gap = 2 mm. FOV = 36–40 cm, No parallel imaging. Use phase sensitive inversion recovery if available (PSIR)	PSIR is more robust and independent of TI time. Optimal for imaging fibrosis. LV epicardial enhancement in the infero-lateral wall has been reported in classic ARVC and in left dominant forms.

Abbreviations: *ARVC* Arrhythmogenic Right Ventricular Cardiomyopathy, *LV* left ventricle, *FOV* field of view, *FSE* fast spin echo, *PSIR* phase sensitive inversion recovery, *RV* right ventricle, *SSFP* steady state free precession, *TE* echo time, *TI* inversion time, *TR* repetition time, *TSE* Turbo spin echo.

CMR Advantages

- Evaluation of RV volumes/function
- Evaluation of RV wall and wall motion abnormalities
- Evaluation of Fatty Infiltration;
- Evaluation of Fibrosis

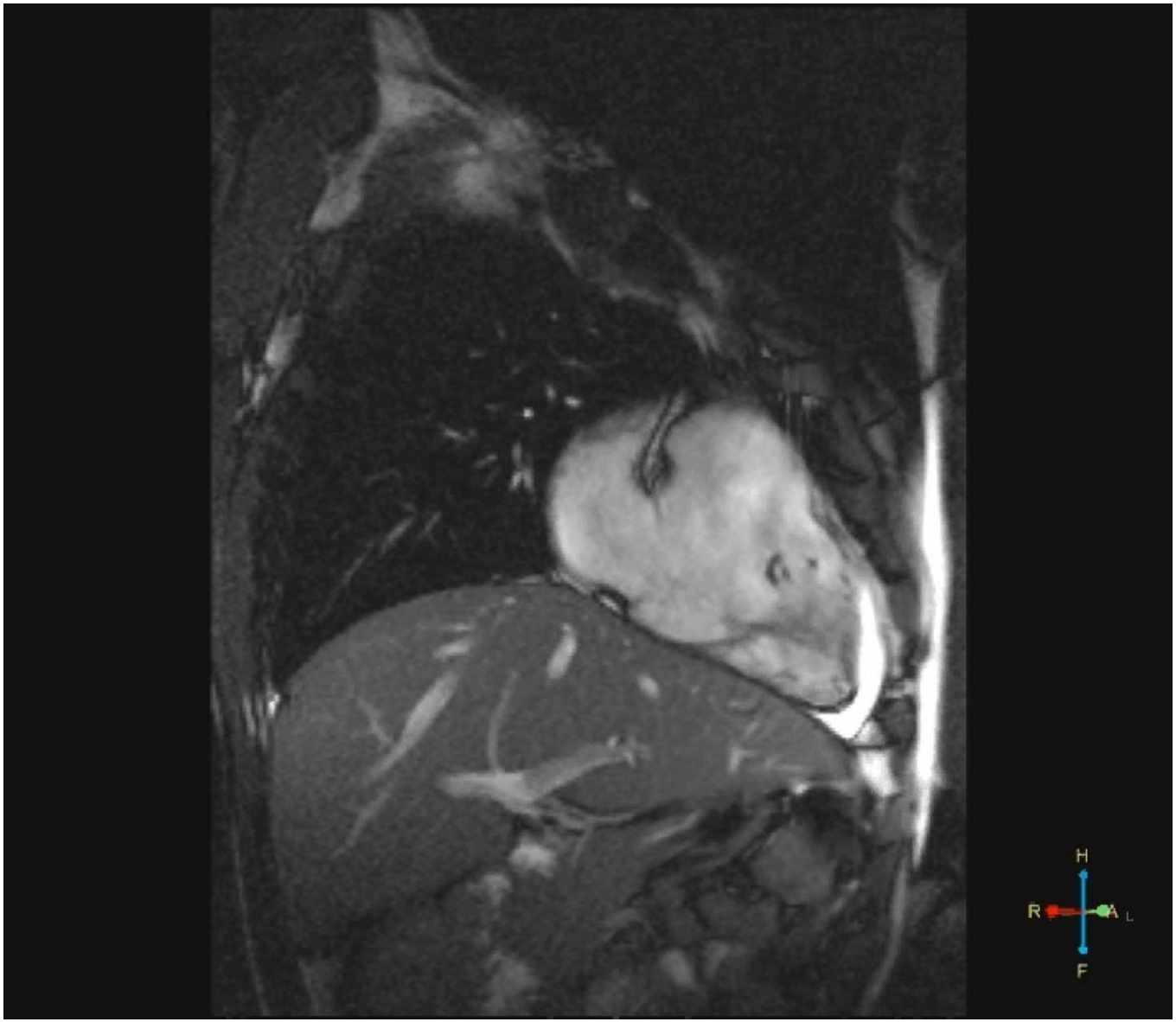
Full-coverage



CMR Advantages

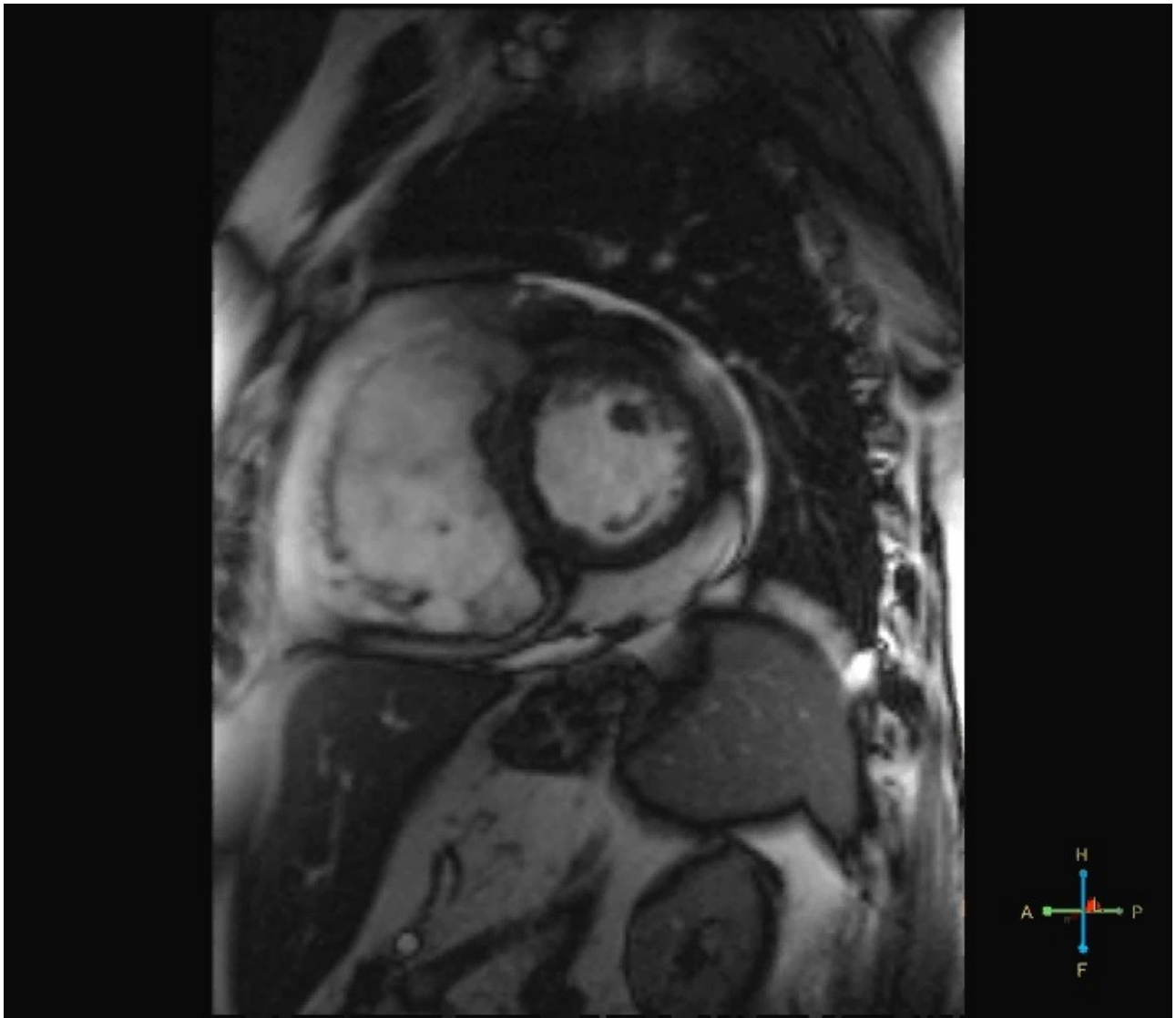
- Evaluation of RV volumes/function
- Evaluation of RV wall and wall motion abnormalities
- Evaluation of Fatty Infiltration;
- Evaluation of Fibrosis

Akinesia



Diskinesia

Abnormal systolic motion (opposite to normal myocardium)

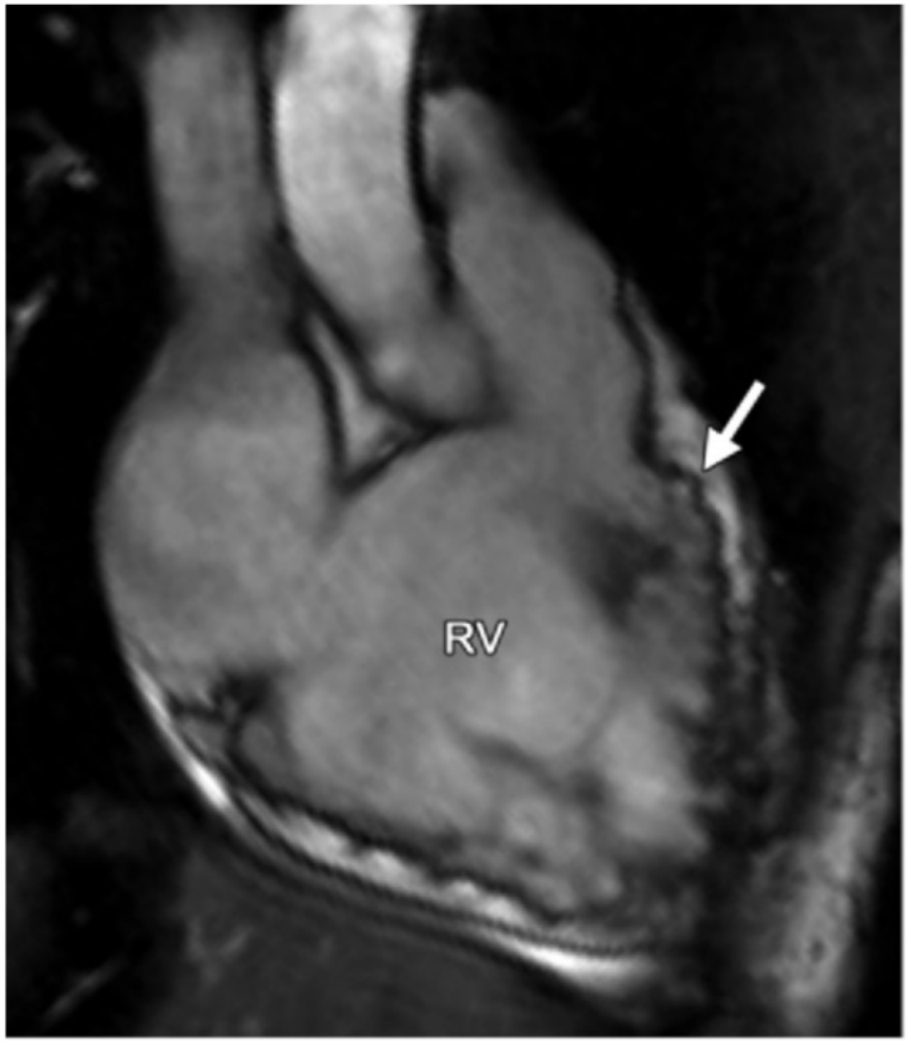


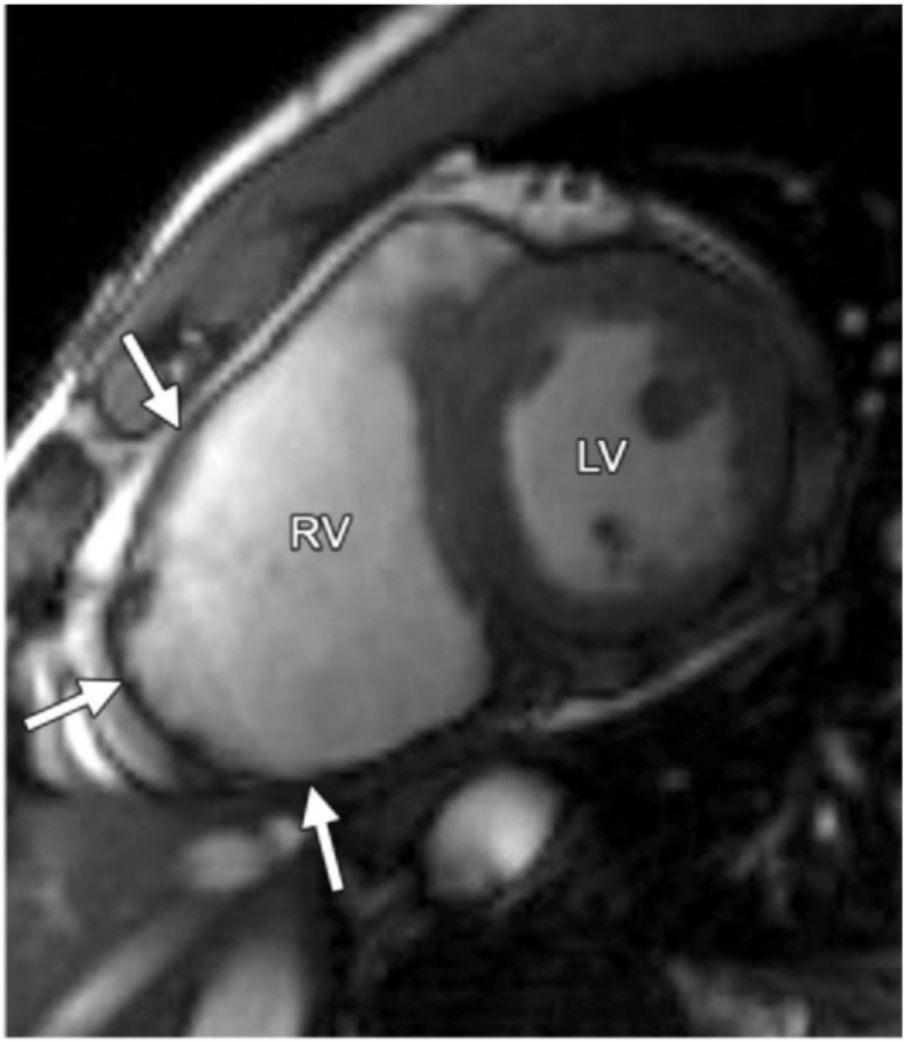
Diskinesia

An akinetic segment with abnormal profile an motion both during systole & diastole.

Tips & Tricks
CINE IMAGES

Guardare alla sede!!!

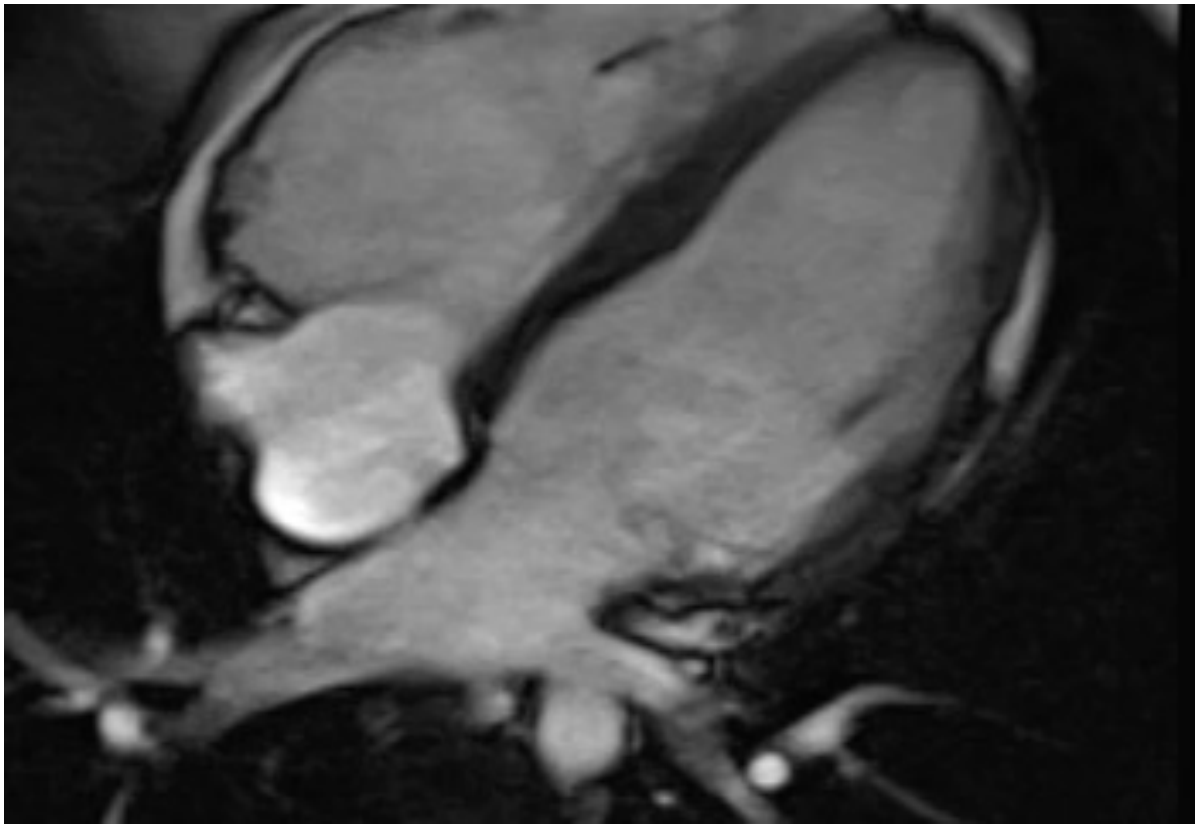




Rastegar N. et al. RadioGraphics 2014; 34:1553–1570

Tips & Tricks
CINE IMAGES

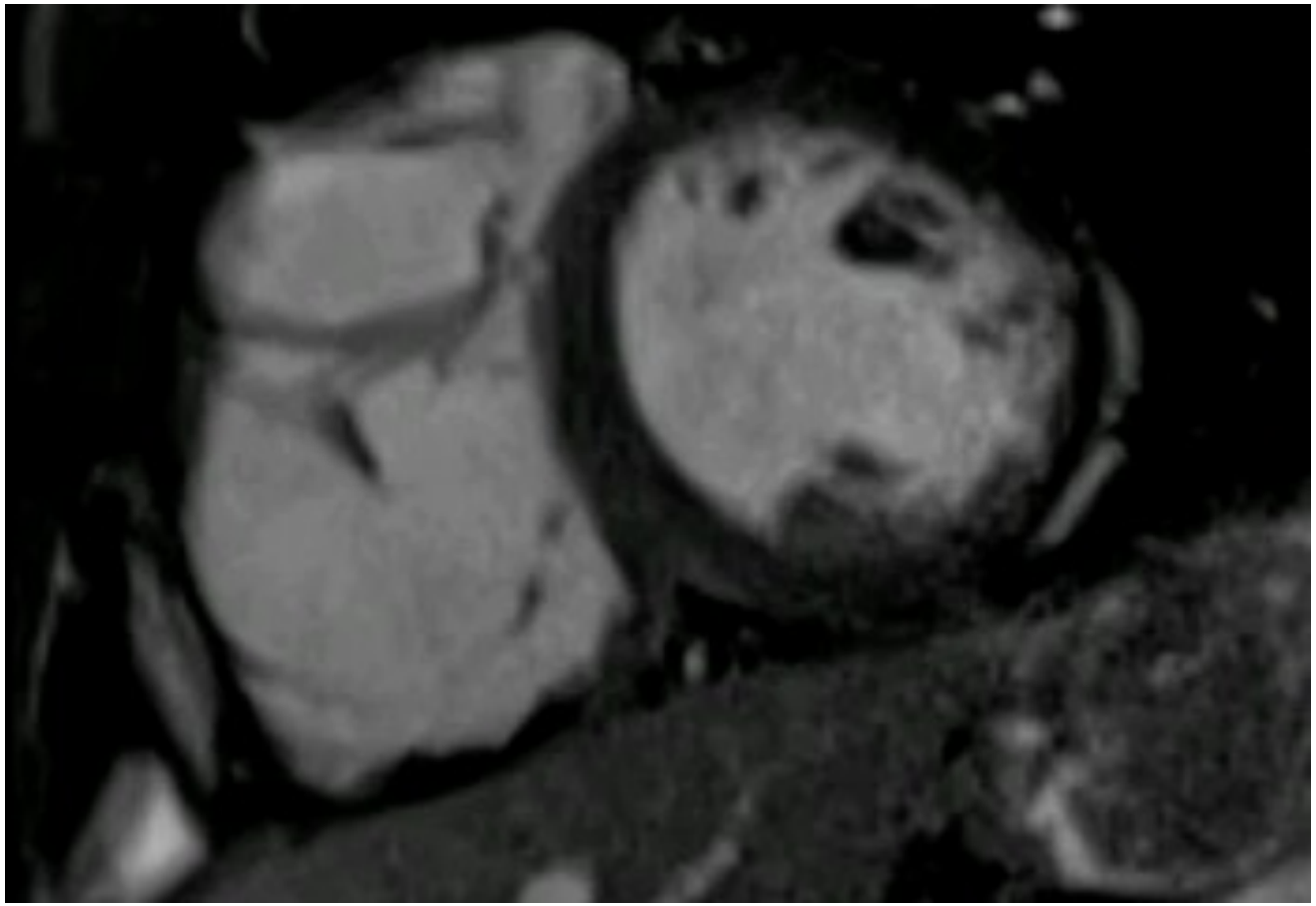
Guardare alla sede!!!



Rastegar N. et al. RadioGraphics 2014; 34:1553–1570

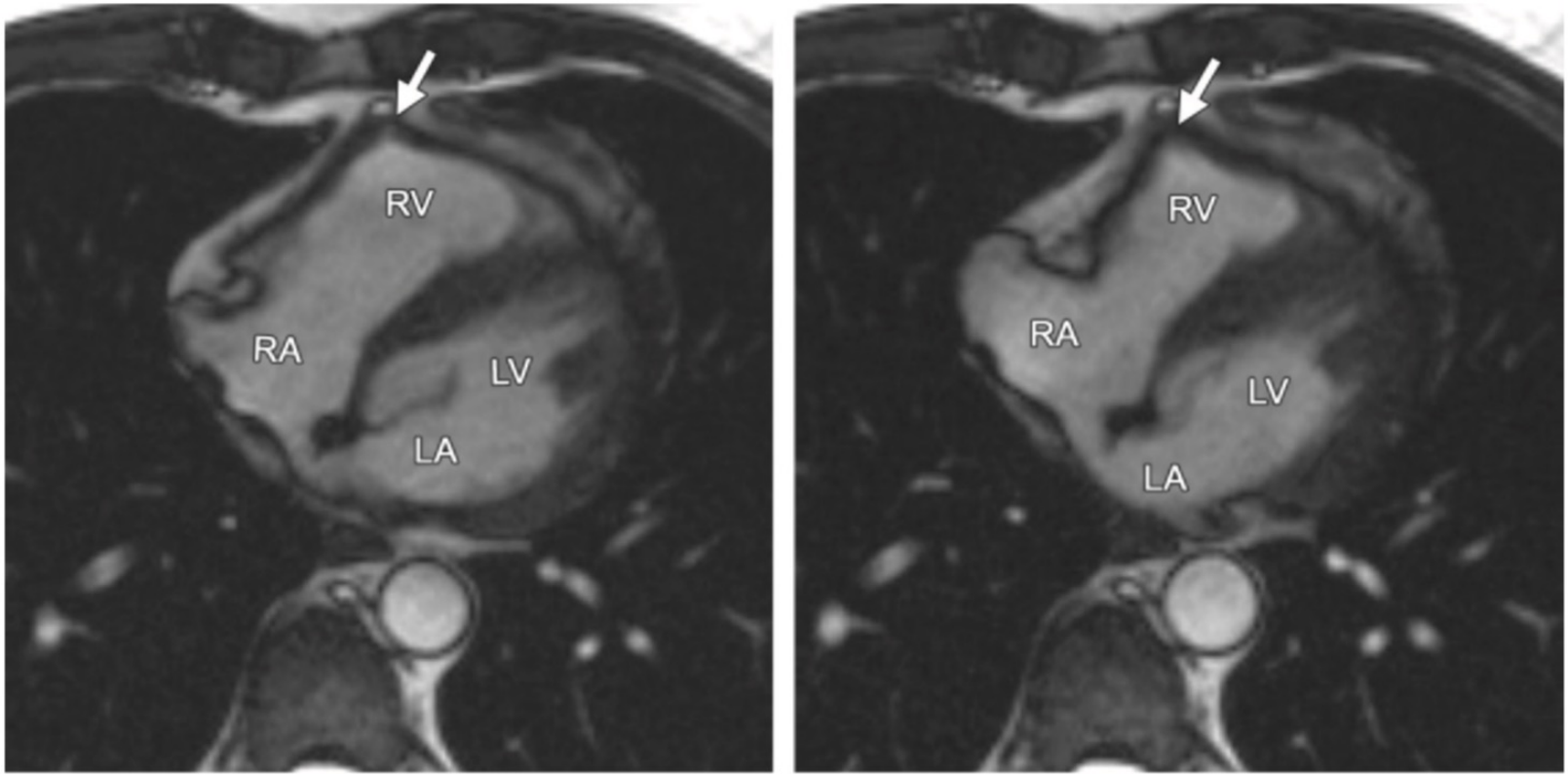
Tips & Tricks
CINE IMAGES

Guardare alla sede!!!



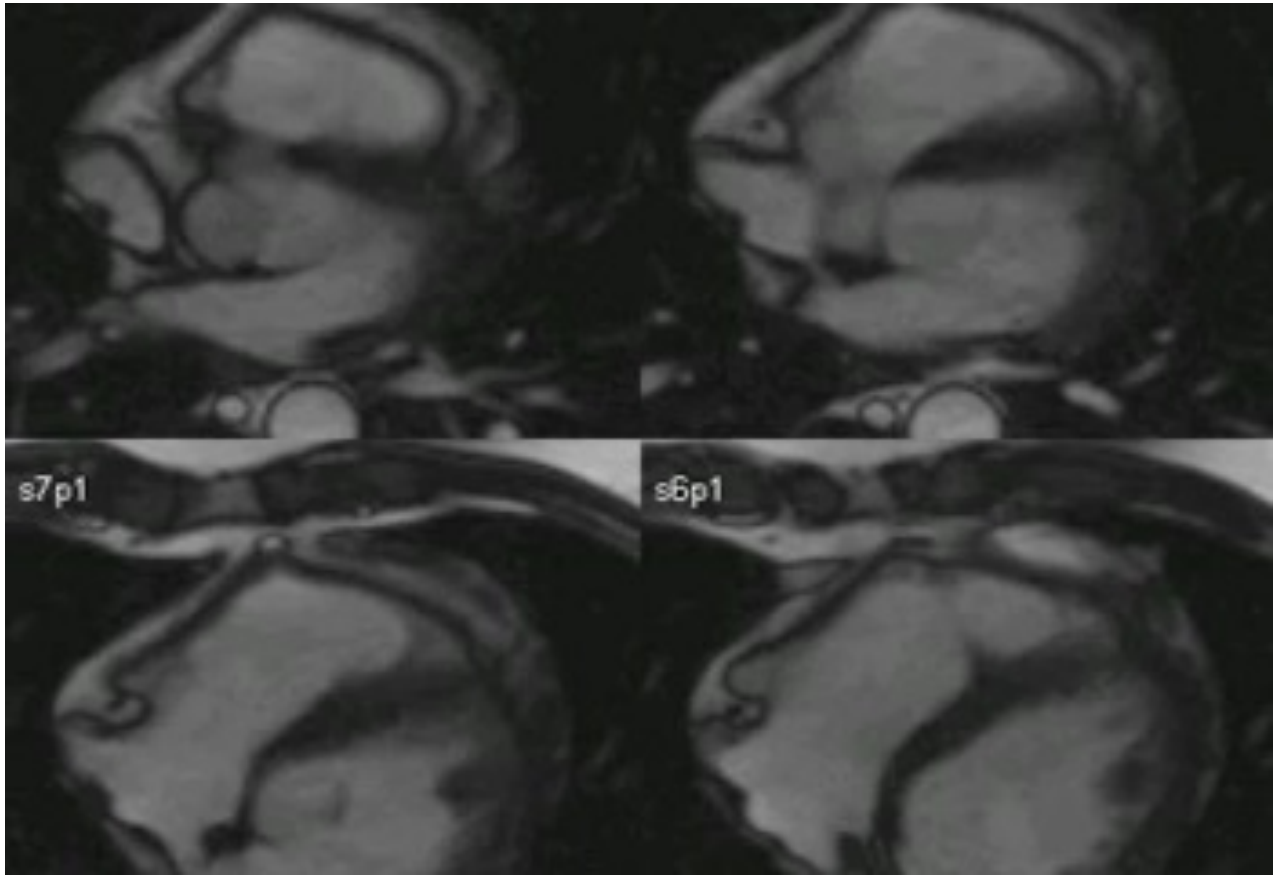
Rastegar N. et al. RadioGraphics 2014; 34:1553–1570

RV Free Wall Tether



Rastegar N. et al. RadioGraphics 2014; 34:1553–1570

RV Free Wall Tether

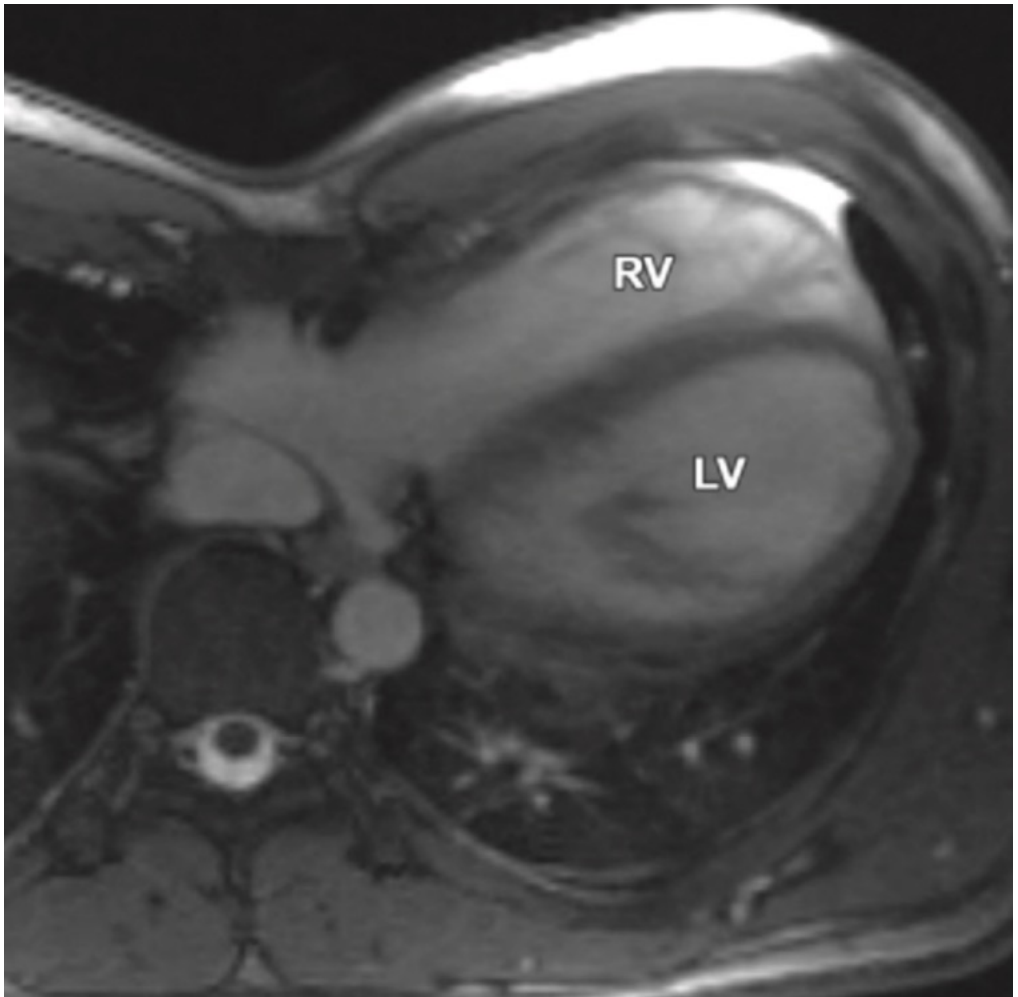


Nelle cine, la porzione «tethered» del VDX (parete libera) rimane nella stessa posizione, immobile, rispetto al resto del miocardio.

Si riesce a vedere la cosiddetta «Triangular-shaped RV» il cui aspetto rimane invariato in sistole.

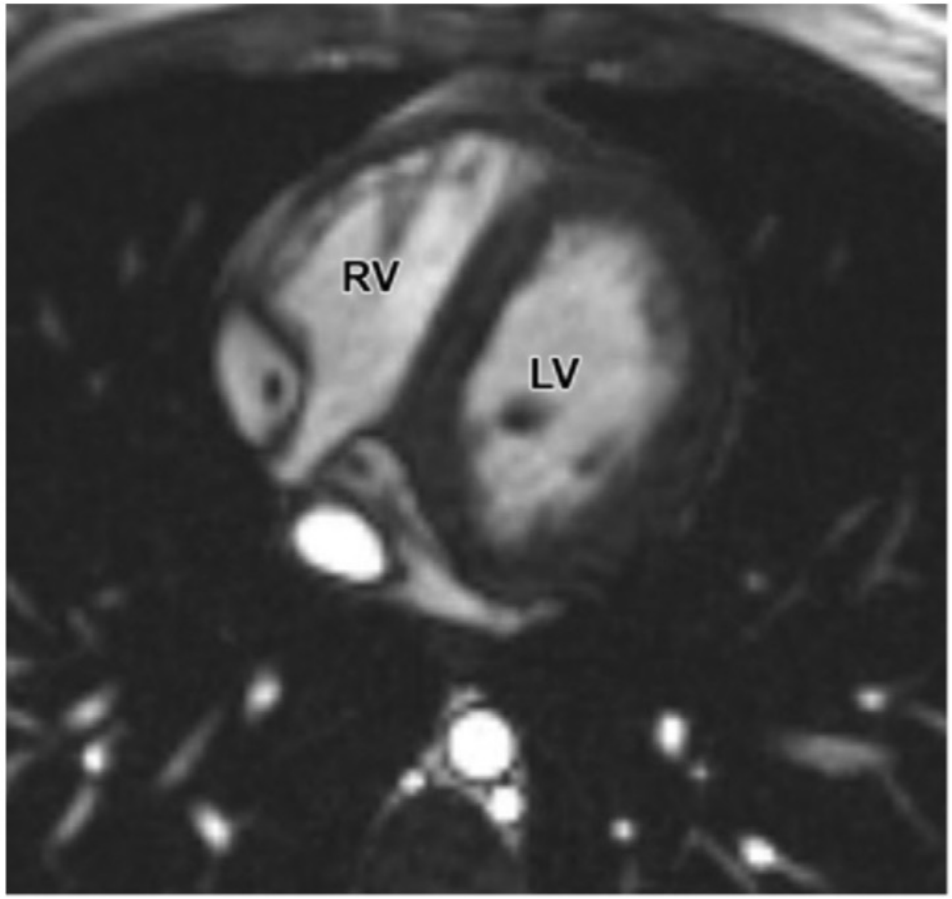
Rastegar N. et al. RadioGraphics 2014; 34:1553–1570

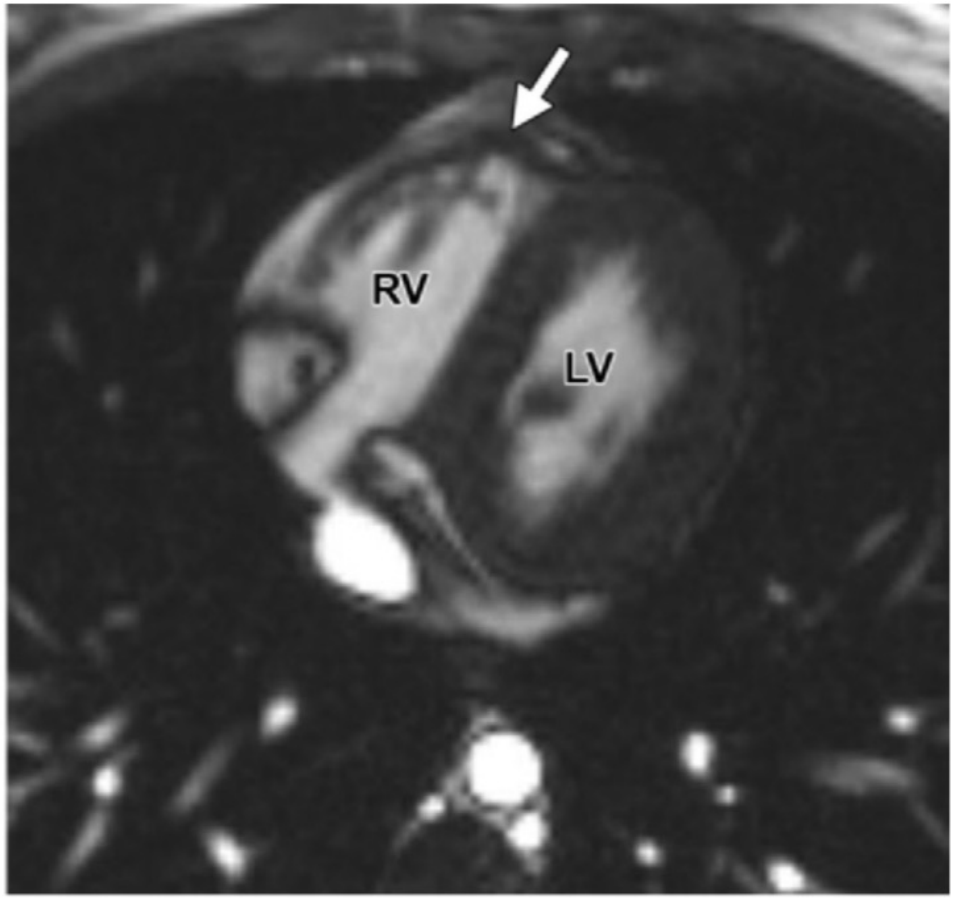
Pectus Excavatum



“banana-shaped RV”

Apicolateral Bulge





Rastegar N. et al. RadioGraphics 2014; 34:1553–1570

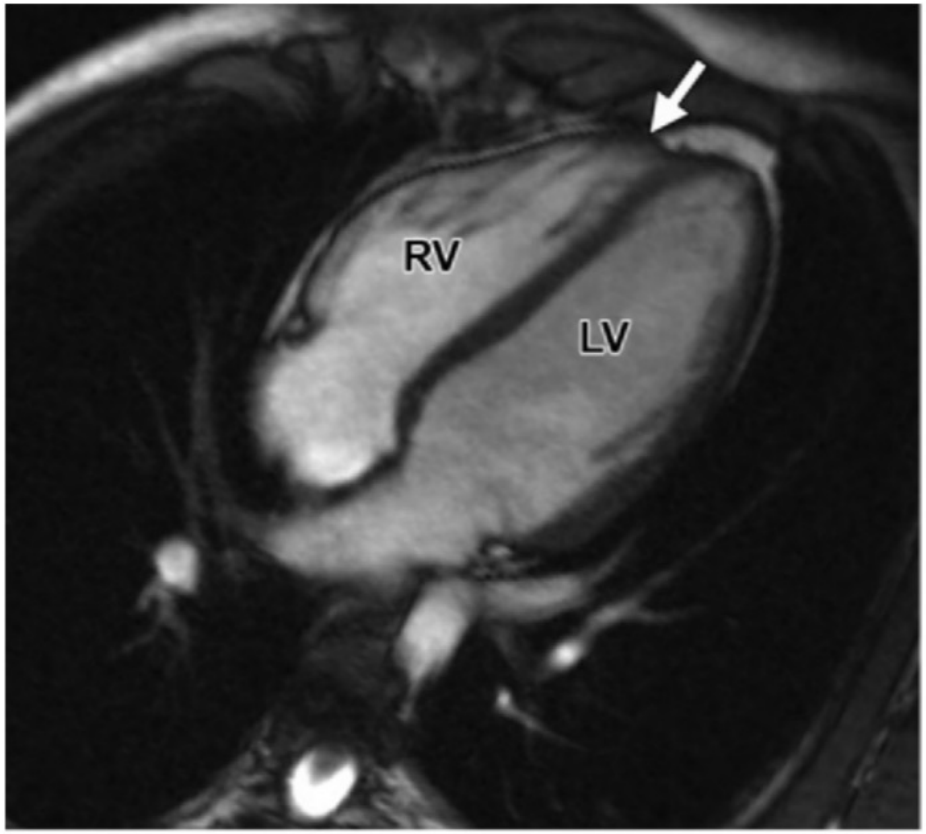
Apicolateral Bulge

At the insertion site of the moderator band in the apicolateral wall

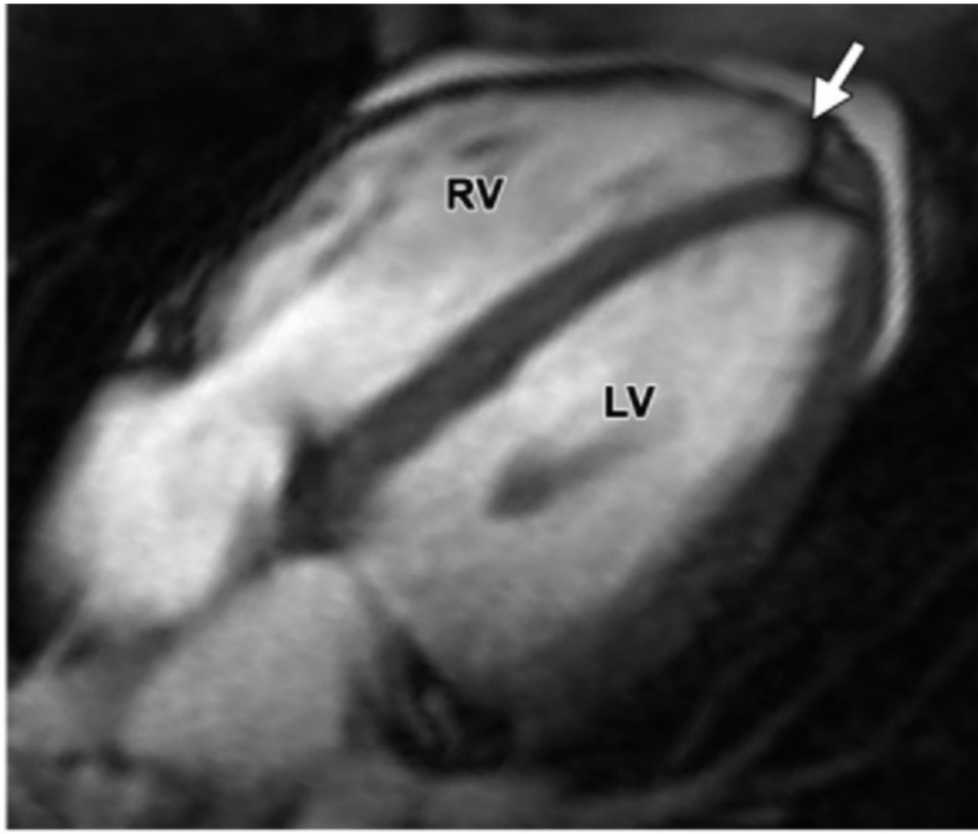
Rastegar N. et al. RadioGraphics 2014; 34:1553–1570

Apical Aneurysm

NORMAL



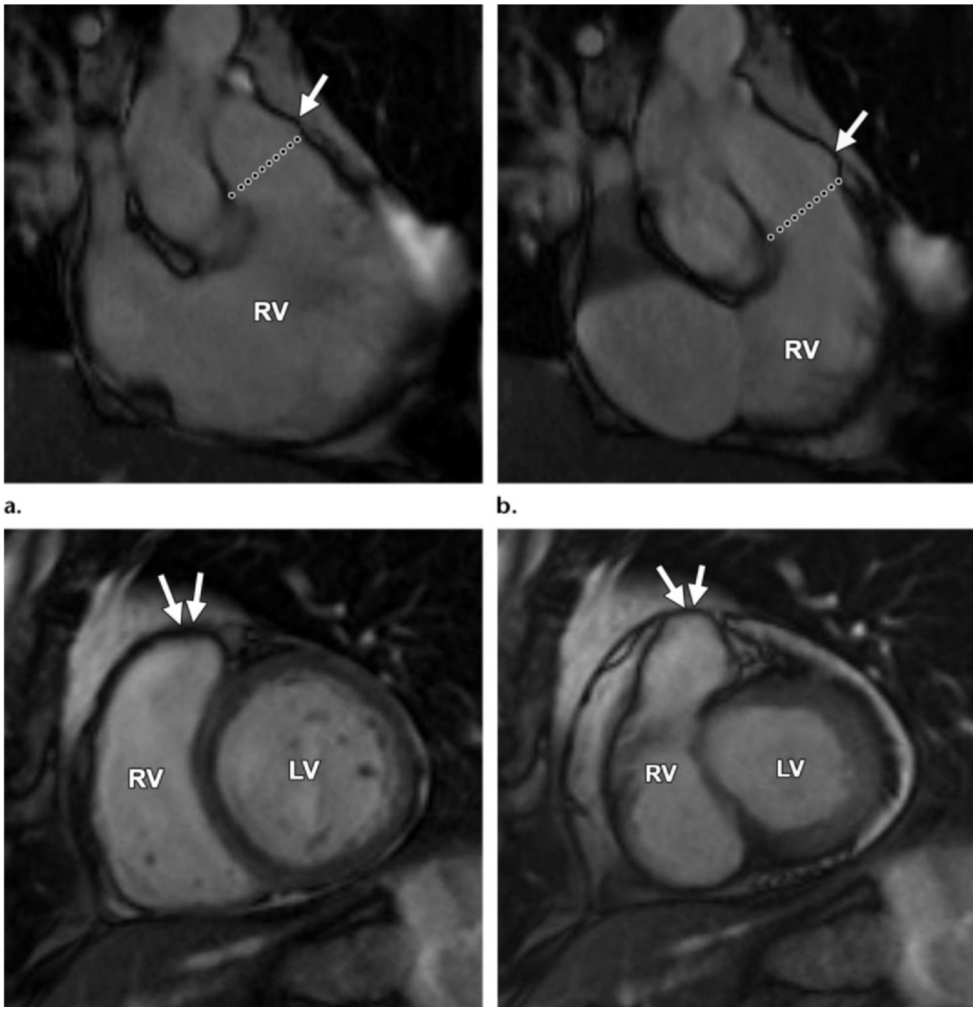
«BUTTERFLY APEX»



Rastegar N. et al. RadioGraphics 2014; 34:1553–1570

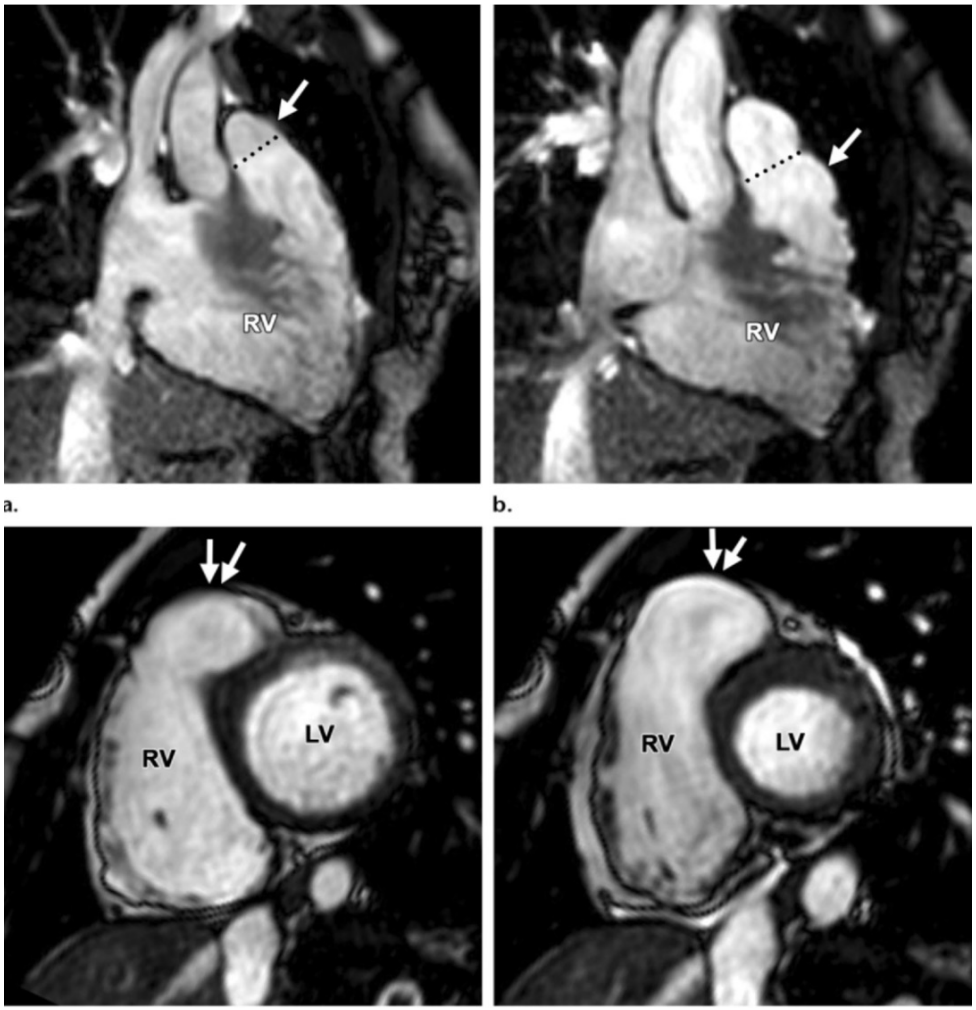
NORMAL

Pulmonary Valve Sinuses



ARVC

Pulmonary Valve Sinuses

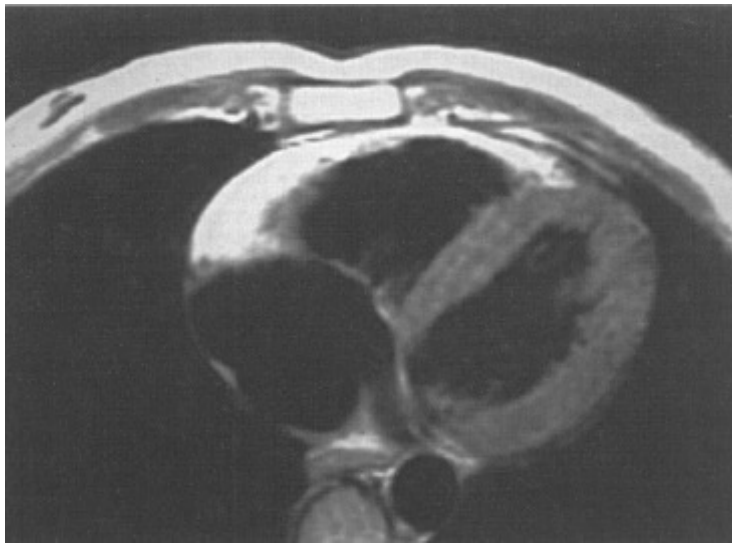


CMR Advantages

- Evaluation of RV volumes/function
- Evaluation of RV wall and wall motion abnormalities
- Evaluation of Fatty Infiltration;
- Evaluation of Fibrosis

The history of tissue characterization by cardiac magnetic resonance imaging

- CMR originally attracted interest in ARVC owing to the ability of fast spin-echo images to differentiate between normal myocardium and fat.
- Initial results were impressive, with reports that T1-weighted spin echo images correctly identified myocardial fat, both in formalin-fixed hearts and in vivo



The history of tissue characterization by cardiac magnetic resonance imaging

MR Imaging of Arrhythmogenic Right Ventricular Cardiomyopathy: Morphologic Findings and Interobserver Reliability

Cardiology 2003;99:153–162

Magnetic Resonance Imaging of Arrhythmogenic Right Ventricular Dysplasia

Sensitivity, Specificity, and Observer Variability of Fat Detection Versus Functional Analysis of the Right Ventricle

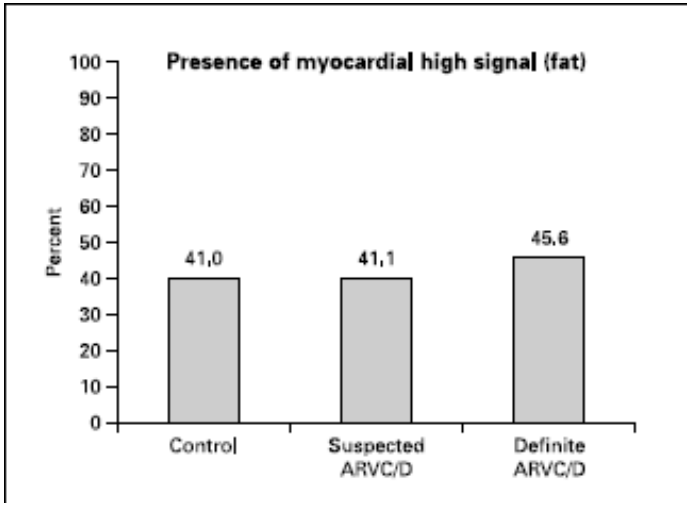
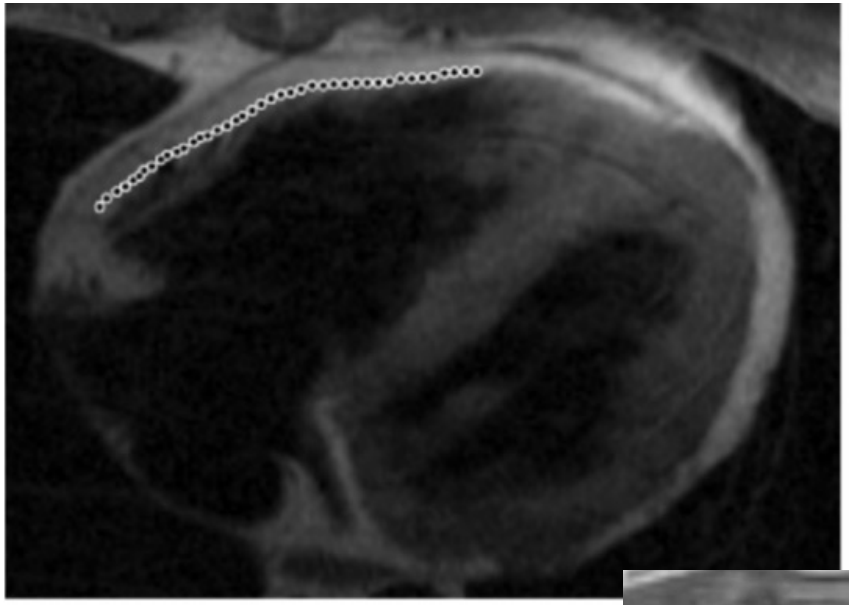


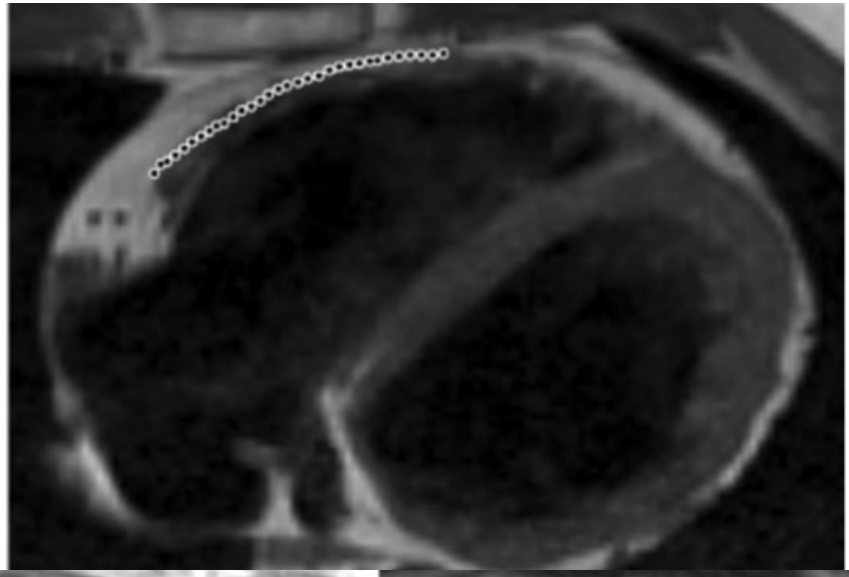
Table 4. Incidence and Interobserver Agreement for the Qualitative Magnetic Resonance Variables

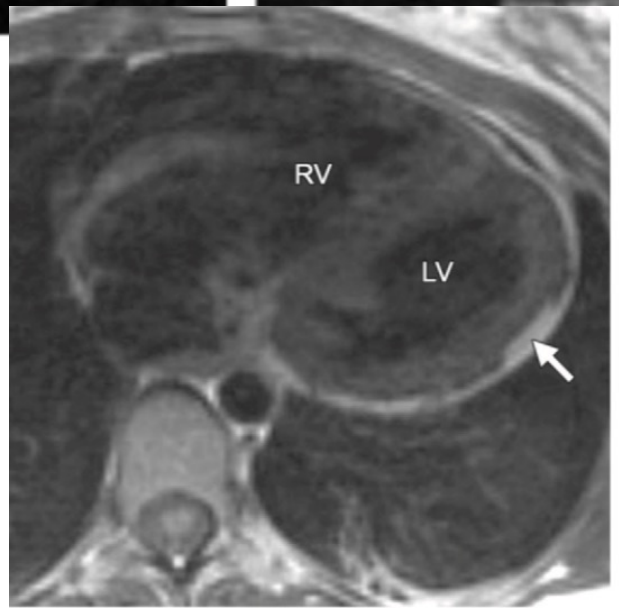
Variable	ARVD	IVT	Controls	% Agreement	Kappa	z-Score	p Value
Intramyocardial fat	84%	34%	4%	88%	0.74	5.75	<0.001
Wall thinning	6%	0%	0%	100%	0.29	2.26	0.01
RV dilation	68%	4%	0%	95%	0.84	6.52	<0.001
Regional function	78%	0%	4%	97%	0.90	6.92	<0.001
RV outflow tract enlargement	66%	0%	4%	93%	0.78	5.13	<0.001

Fat infiltration of the RV had a sensitivity of 84% and specificity of 79% with a positive predictive value of 80%. Regional wall motion abnormality of the RV had a sensitivity of 75% and specificity of 97% with a positive predictive value of 90%.

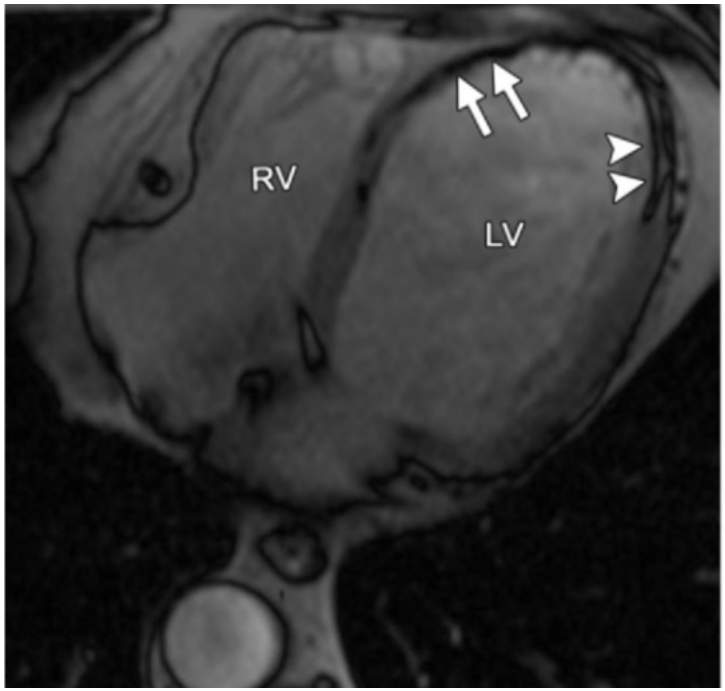
Myocardial FAT in ARVC

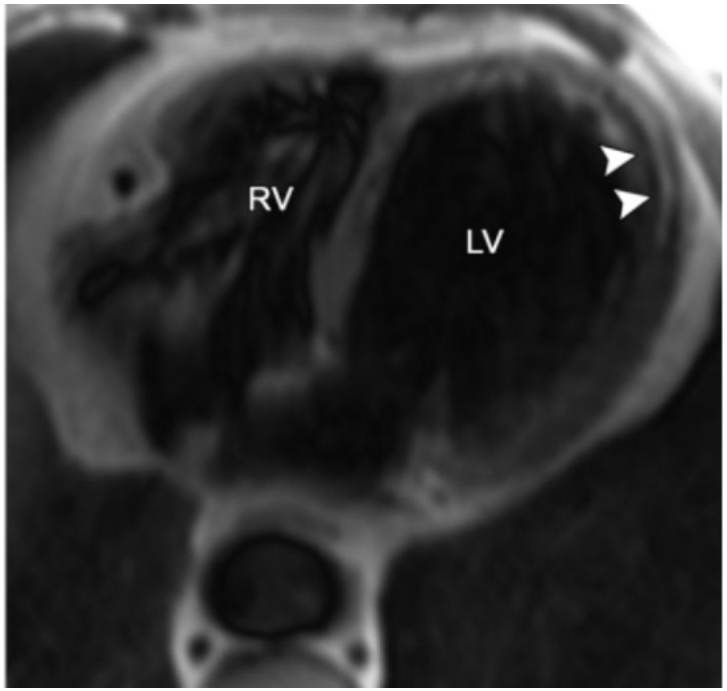






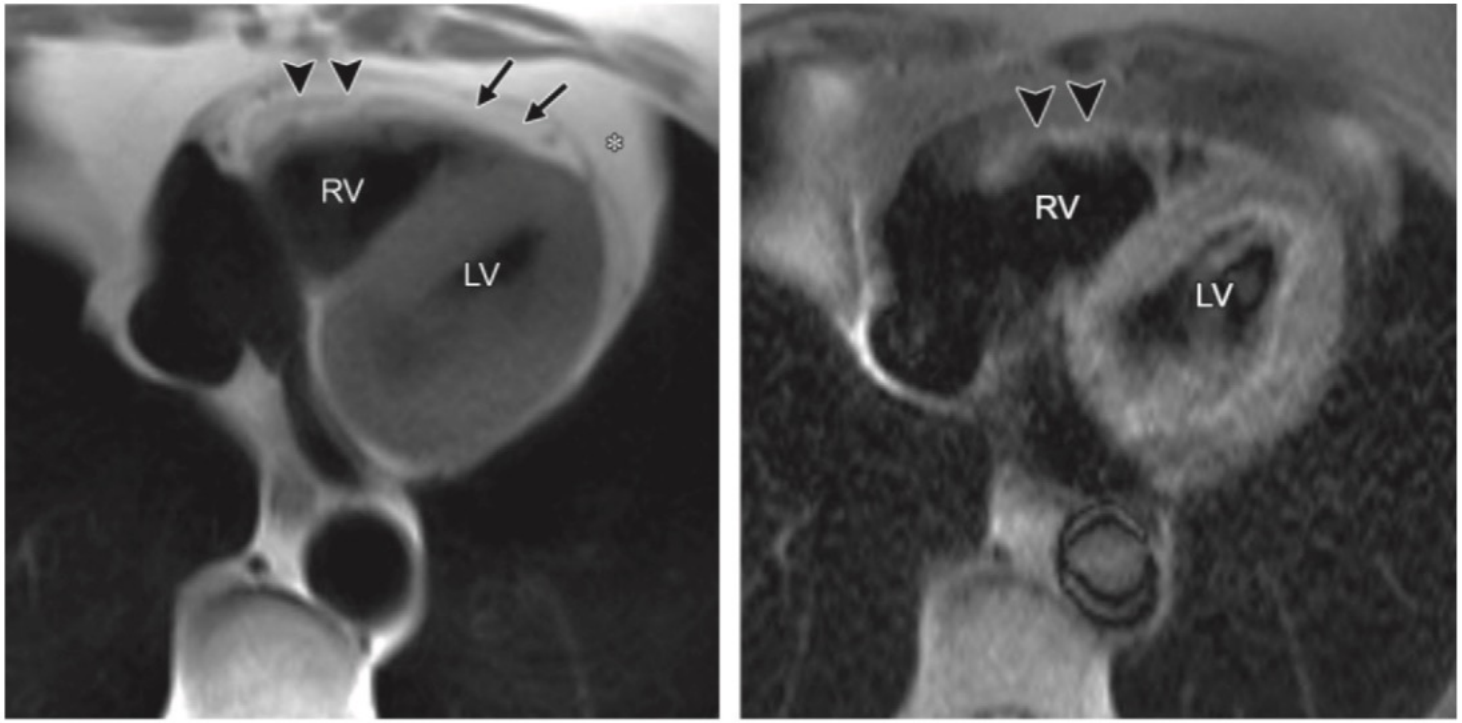
Myocardial FAT in other conditions





CICATRICE POST-INFARTUALE

Myocardial FAT in other conditions



ADIPOSITAS CORDIS

Tips & Tricks
CINE IMAGES

1. Evaluation of fibrofatty infiltration only as a secondary assessment after the evaluation of the more reliable functional and structural assessment of the RV;

2. Compare with cine images

Just a matter of fat?

Basso C Cardiovasc Pathology 2005;14:37-41

"ARVC is predominantly a genetically determined heart muscle disorder that is characterized pathologically by fibrofatty replacement of the right ventricular myocardium"

Marcus et al. EHJ 2010;31, 806–814

Is there a chance for fibrosis detection by CMR?

Tissue characterization of wall modified Task Force 2010

Major

- 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

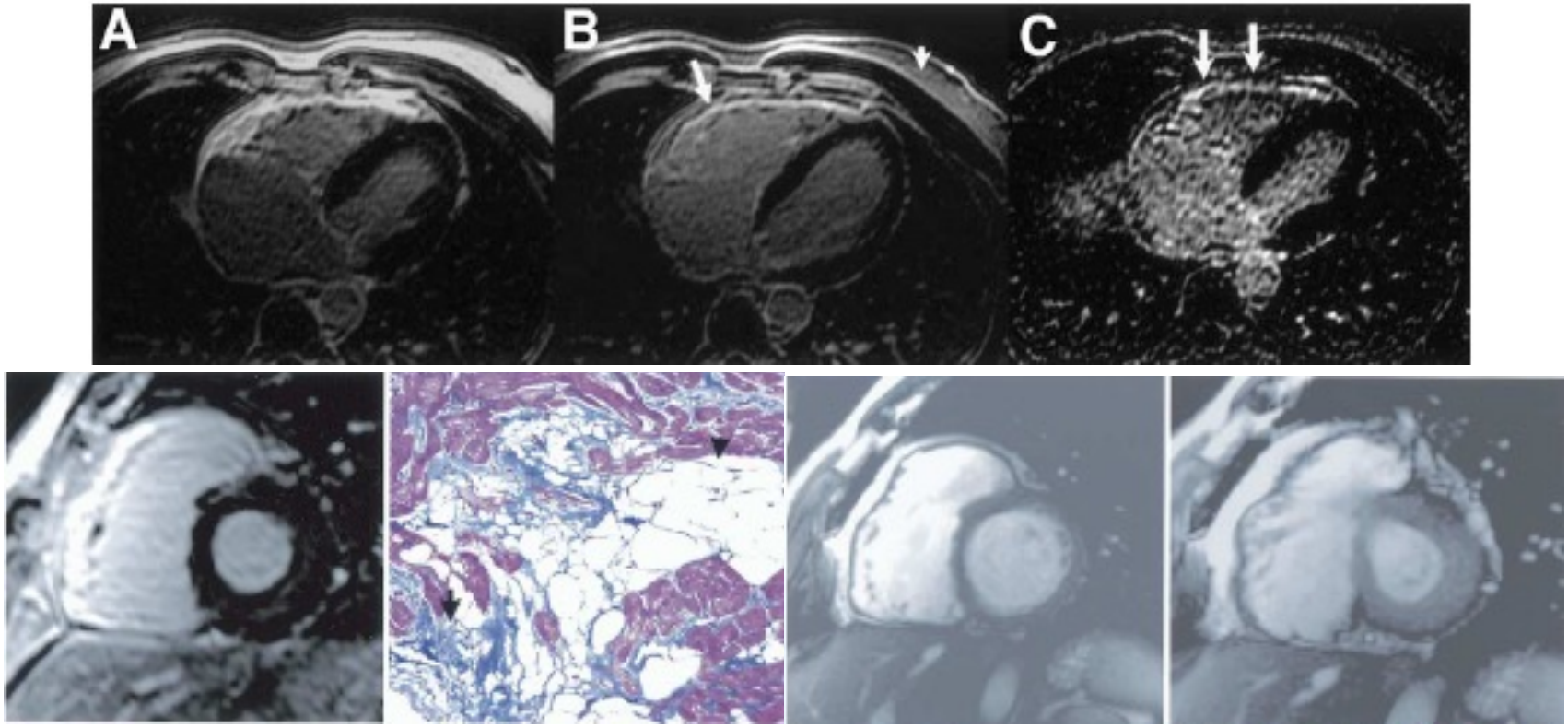
- Residual myocytes 60% to 75% by morphometric analysis (or 50% to 65% if estimated), with fibrous replacement of the RV free wall myocardium in ≥ 1 sample, with or without fatty replacement of tissue on endomyocardial biopsy

CMR Advantages

- Evaluation of RV volumes/function
- Evaluation of RV wall and wall motion abnormalities
- Evaluation of Fatty Infiltration;
- Evaluation of Fibrosis

Noninvasive Detection of Myocardial Fibrosis in Arrhythmogenic Right Ventricular Cardiomyopathy Using Delayed-Enhancement Magnetic Resonance Imaging

Tandri H et al J Am Coll Cardiol 2005;45:98 –103



Eight (67%) of the 12 ARVC patients demonstrated LGE in the RV compared with none (0%) of the 18 patients without ARVC ($p < 0.001$).

Noninvasive Detection of Myocardial Fibrosis in Arrhythmogenic Right Ventricular Cardiomyopathy Using Delayed-Enhancement Magnetic Resonance Imaging

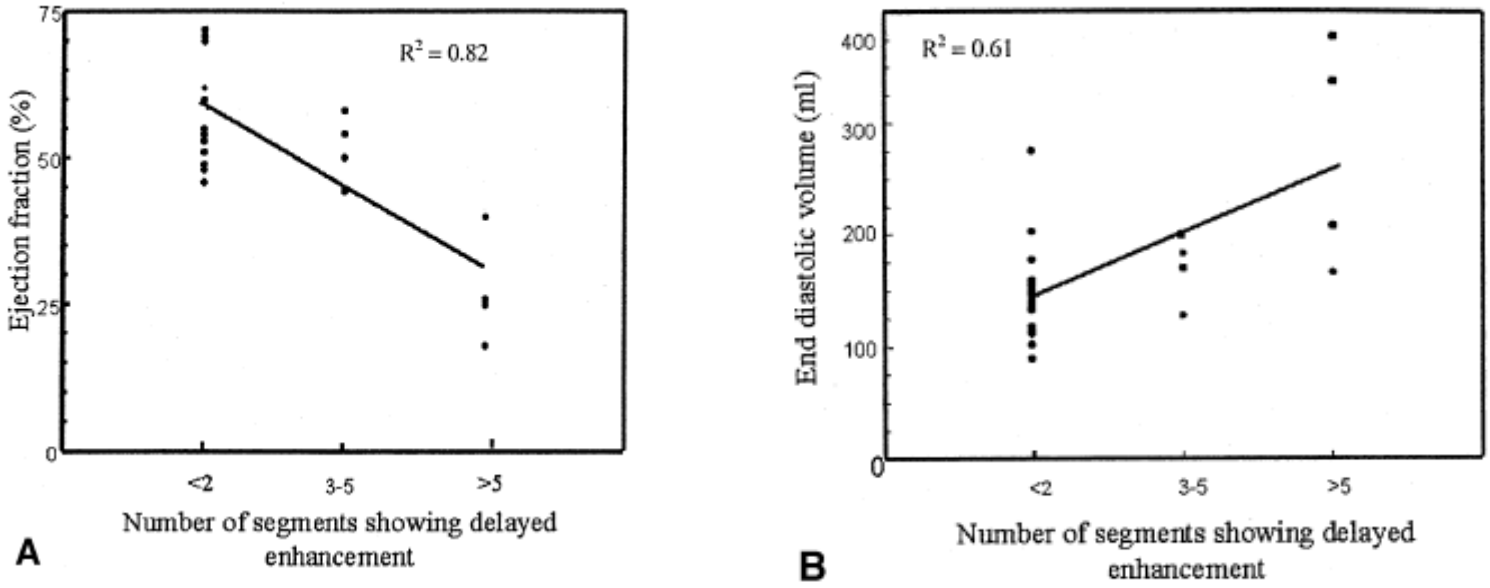
Table 2. Correlation of MR Myocardial Delayed Enhancement With EP and Histologic Findings in ARVD Patients

	MR Imaging				EP Testing			Histopathology		
Case	Fat	Regional Function	MDE	Segments Abnormal	Arrhythmia Morphology	Site	Length (ms)	Fat	Fibrosis	Inflammation
1	ant/RVOT	ant-DK	ant	7	LBBB-SA	apex	260	+	+++	none
2	none	ant-DK	ant	4	RBBB-LA	apex	202	+	+++	none
3	ant/RVOT	ant-DK	diffuse	6	LBBB-IA	RVOT	240	+	+++	+
4	basal	normal	none	0	noninducible	—	—	none	none	none
5	basal	basal DK	basal	3	noninducible	—	—	ND	ND	ND
6	none	RVOT DK	none	0	not done	—	—	ND	ND	ND
7	none	normal	none	0	noninducible	—	—	none	none	none
8	ant/RVOT	basal DK	ant-basal	3	LBBB-IA, LBBB-RA	apex	240,232	none	none	none
9	none	normal	none	0	noninducible	—	—	none	none	+
10	none	ant-DK	ant-basal	1	not done	—	—	ND	ND	ND
11	ant/basal	ant-DK	ant/IVS*	8	LBBB-SA	RVOT	230	none	+	+++
12	ant/RVOT	ant-DK	ant	7	RBBB-LA	RVOT	300	+	+++	+

- **LGE correlated better than abnormal signal-averaged ECG (5 of 6) or RV dysfunction (4 of 6) in predicting VT on EP testing.**
- **None of the ARVD/C patients without delayed enhancement was inducible.**

Tandri H et al J Am Coll Cardiol 2005;45:98 –103

Noninvasive Detection of Myocardial Fibrosis in Arrhythmogenic Right Ventricular Cardiomyopathy Using Delayed-Enhancement Magnetic Resonance Imaging



- **The extent of fibrosis showed an inverse correlation with global RV function and a direct correlation with increased RV end-diastolic volume**

Tandri H et al J Am Coll Cardiol 2005;45:98 –103

LGE in ARVC: Controversies and Caveats

- Selection bias-patient population studied (probands vs family members, different genotype, etc);
- Histological correlations;
- Look at LV;
- Differential diagnosis;
- Comparison with kinetics;
- A role in the Task Force Criteria: Padua Criteria

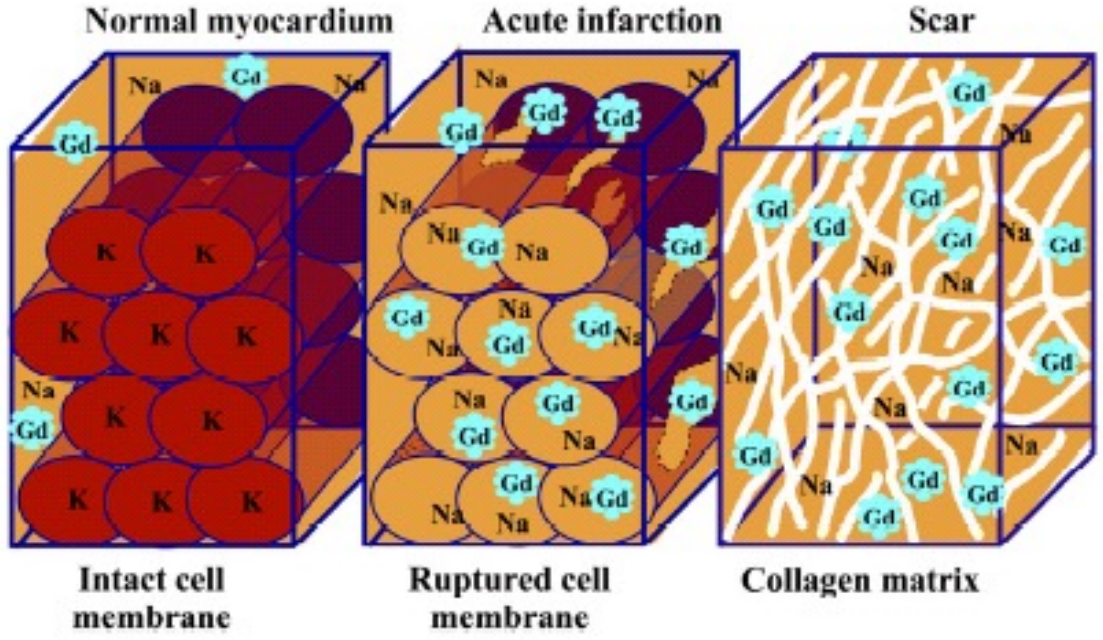
Noninvasive Detection of Myocardial Fibrosis in Arrhythmogenic Right Ventricular Cardiomyopathy Using Delayed-Enhancement Magnetic Resonance Imaging

- **Endomyocardial biopsy was performed in 9 of the 12 patients who had the final diagnosis of ARVD/C.**
  - **4 (44%) fibro-fatty ARVD/C -> all LGE**
  - **1 (11%) myocarditis -> LGE**
  - **4 (44%) had normal biopsies -> only 1 had LGE**

However a definitive histologic-proven basis of RV LGE in ARVC remains to be established

Tandri H et al J Am Coll Cardiol 2005;45:98 –103

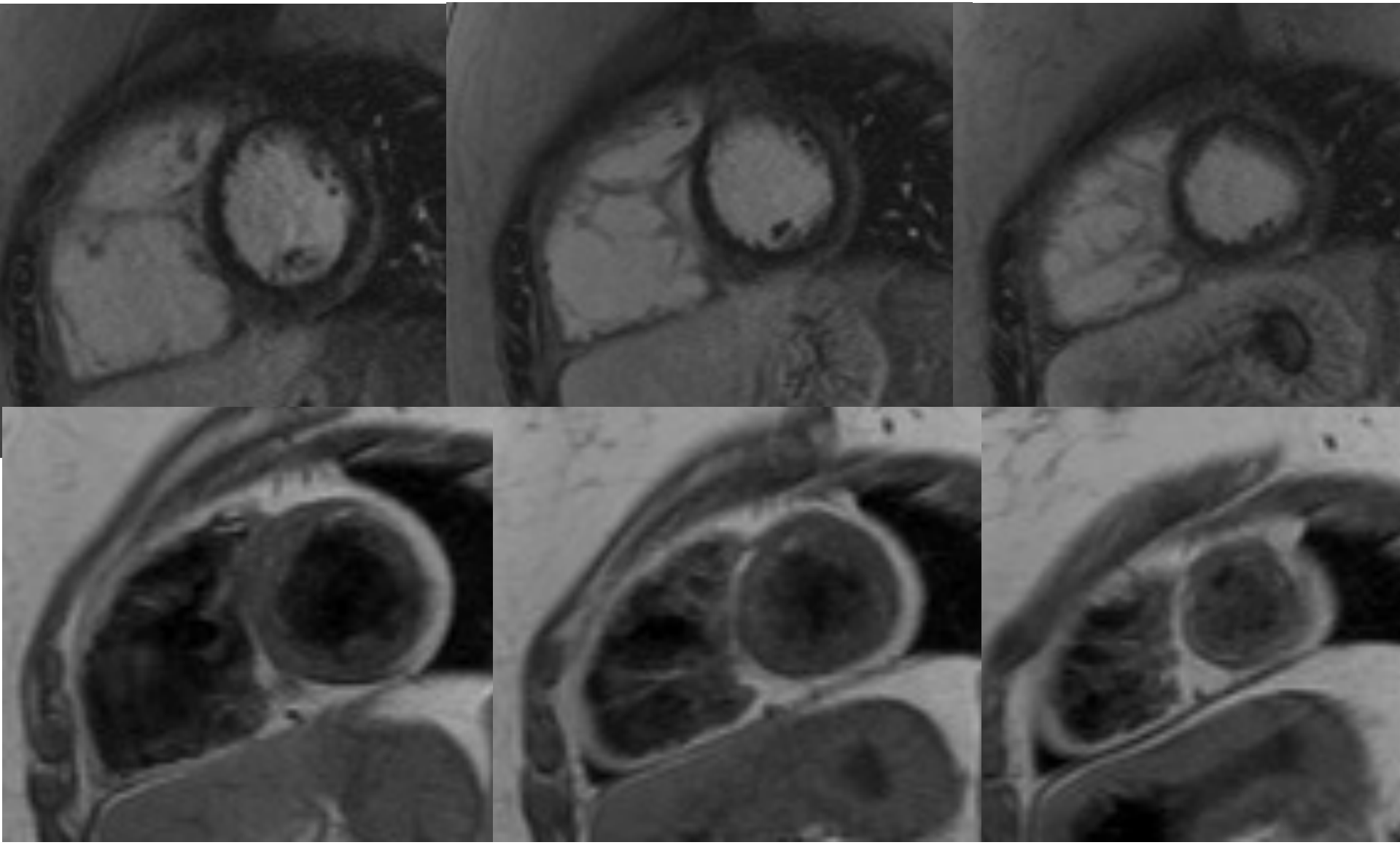
CARATTERIZZAZIONE TISSUTALE & RM



Circulation. 1995;92:1117-1125

CARATTERIZZAZIONE TISSUTALE & RM:

CASO 1



CARATTERIZZAZIONE TISSUTALE & RM:

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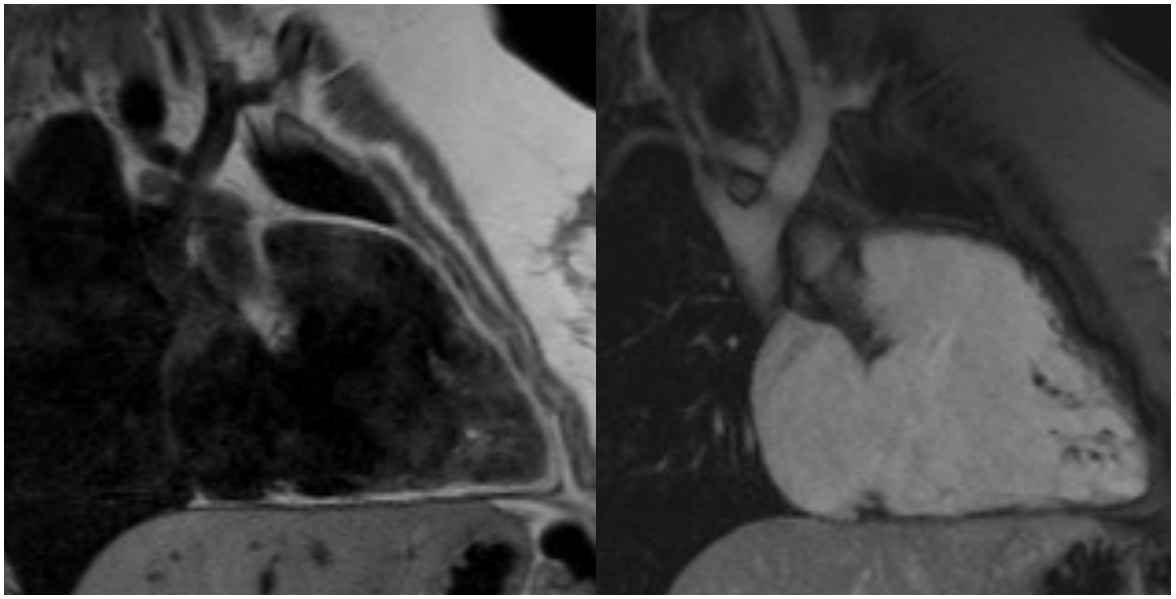

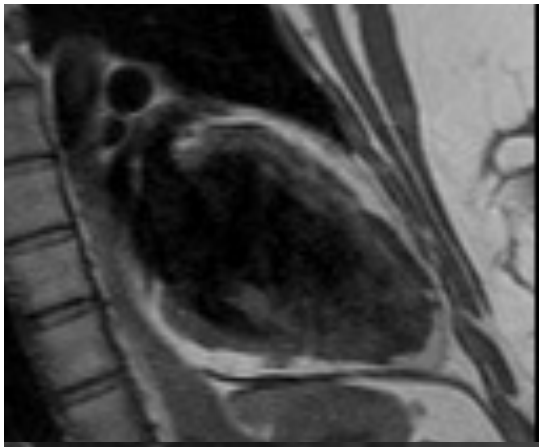

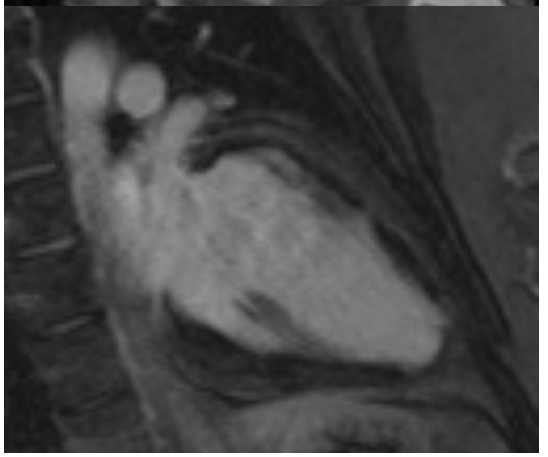

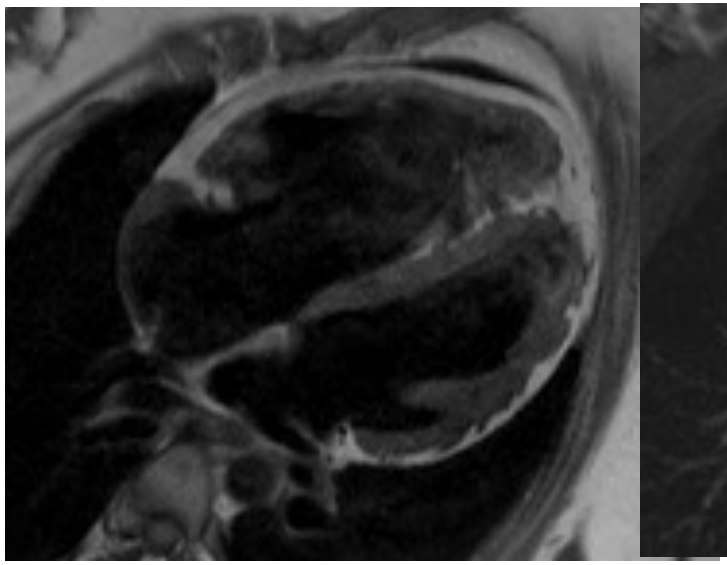

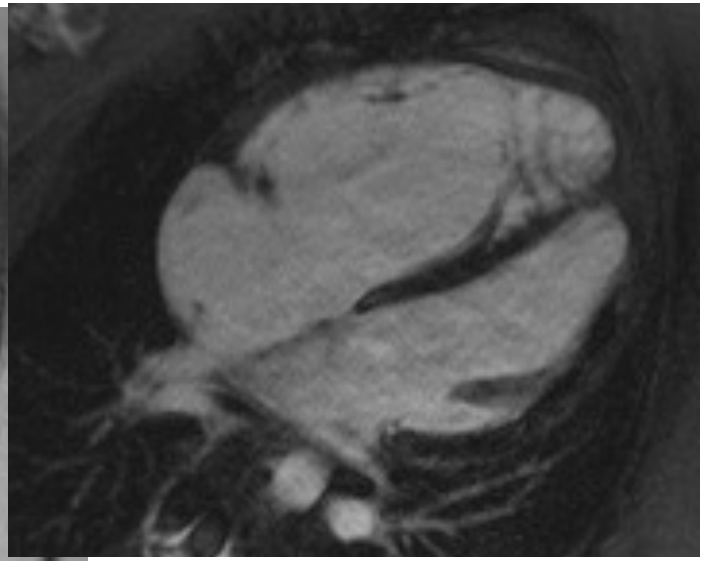

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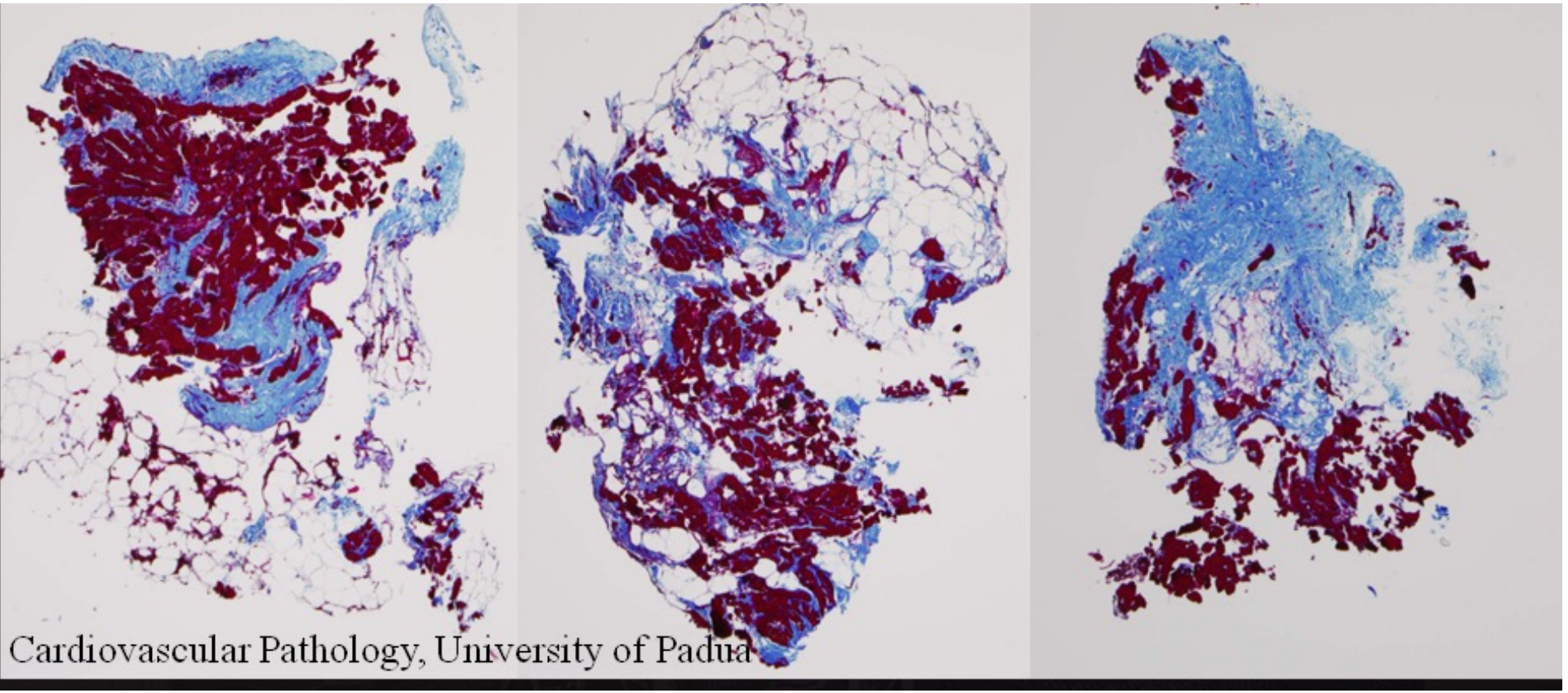
CASO 1



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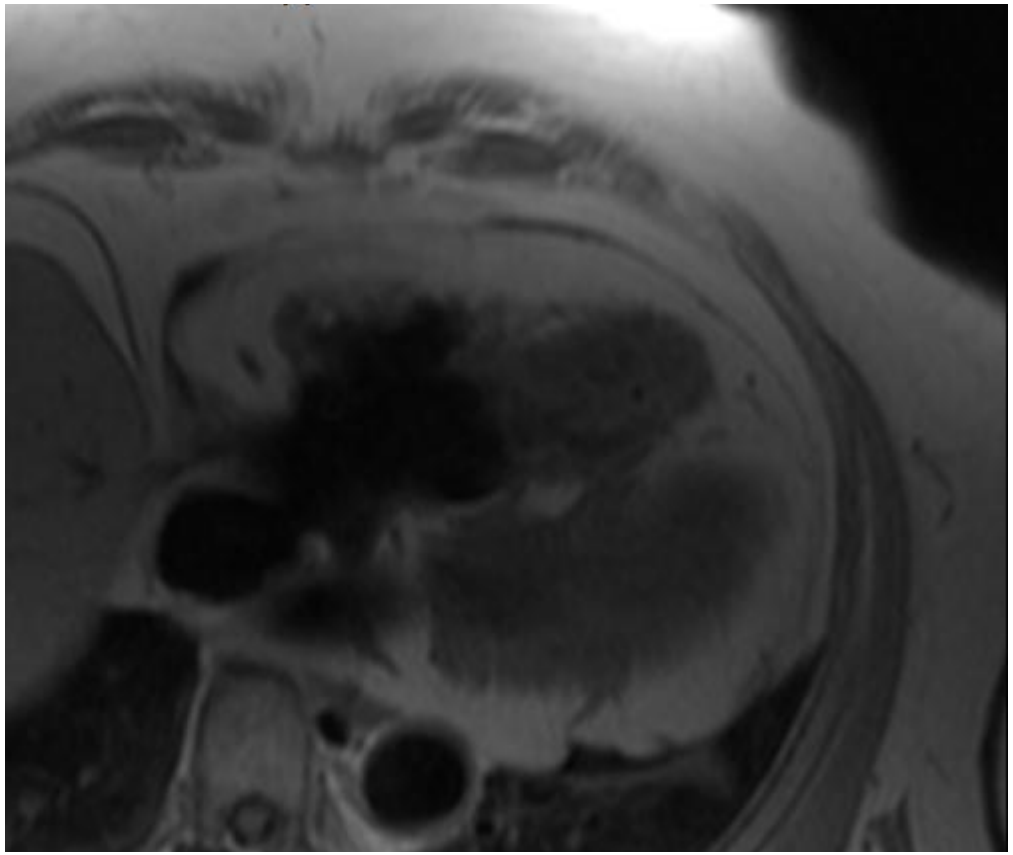

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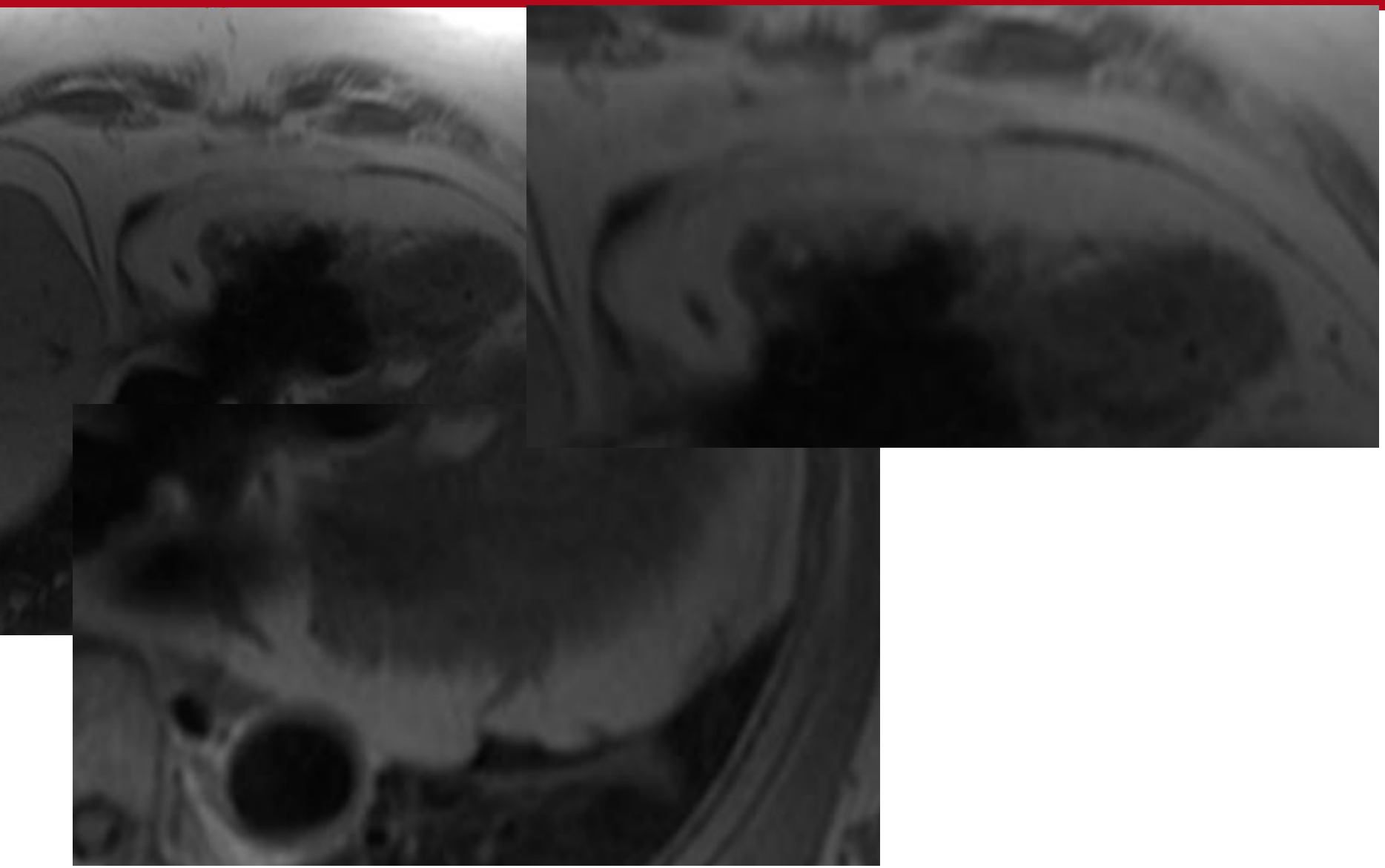

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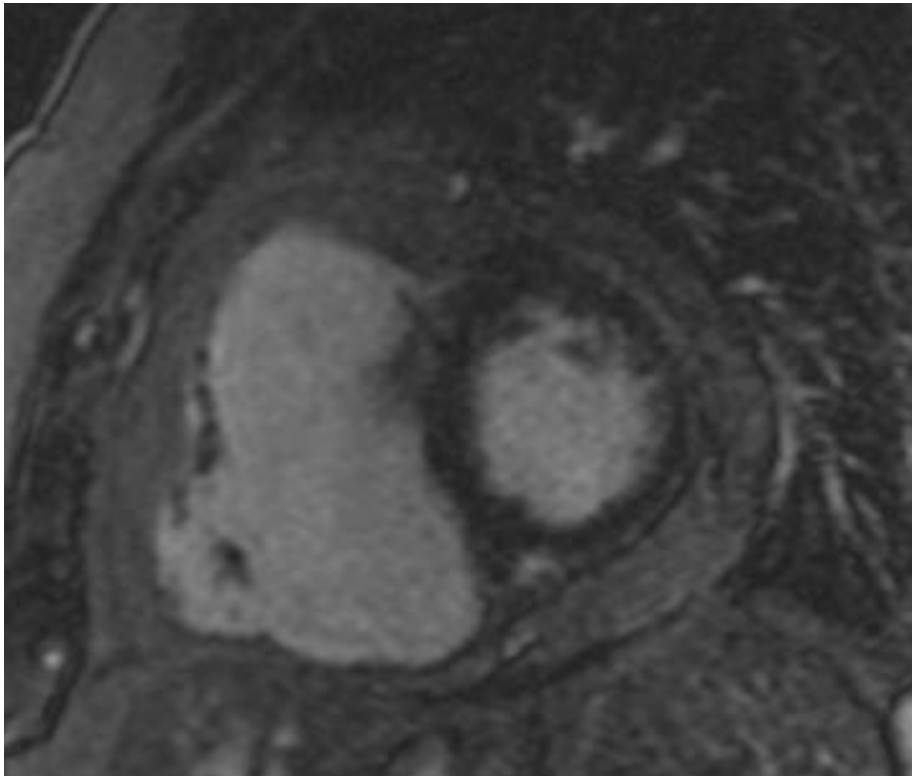

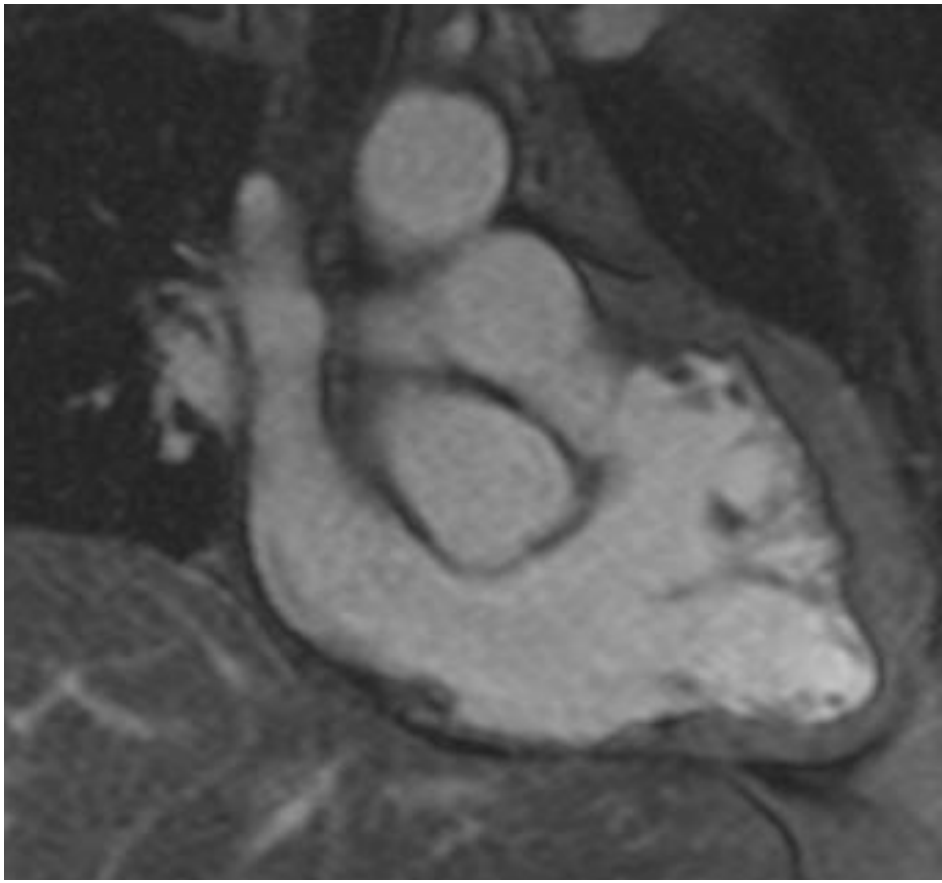

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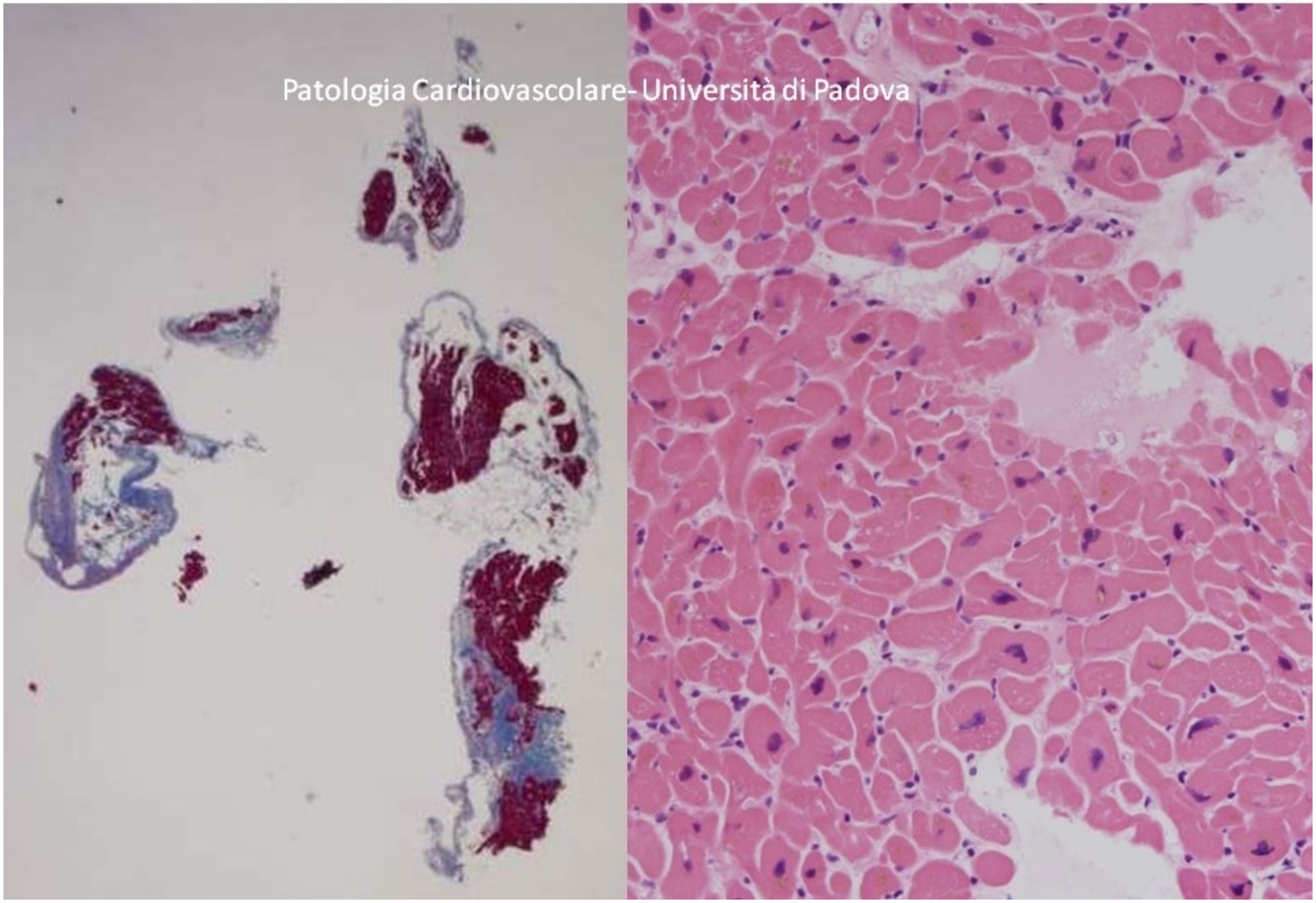

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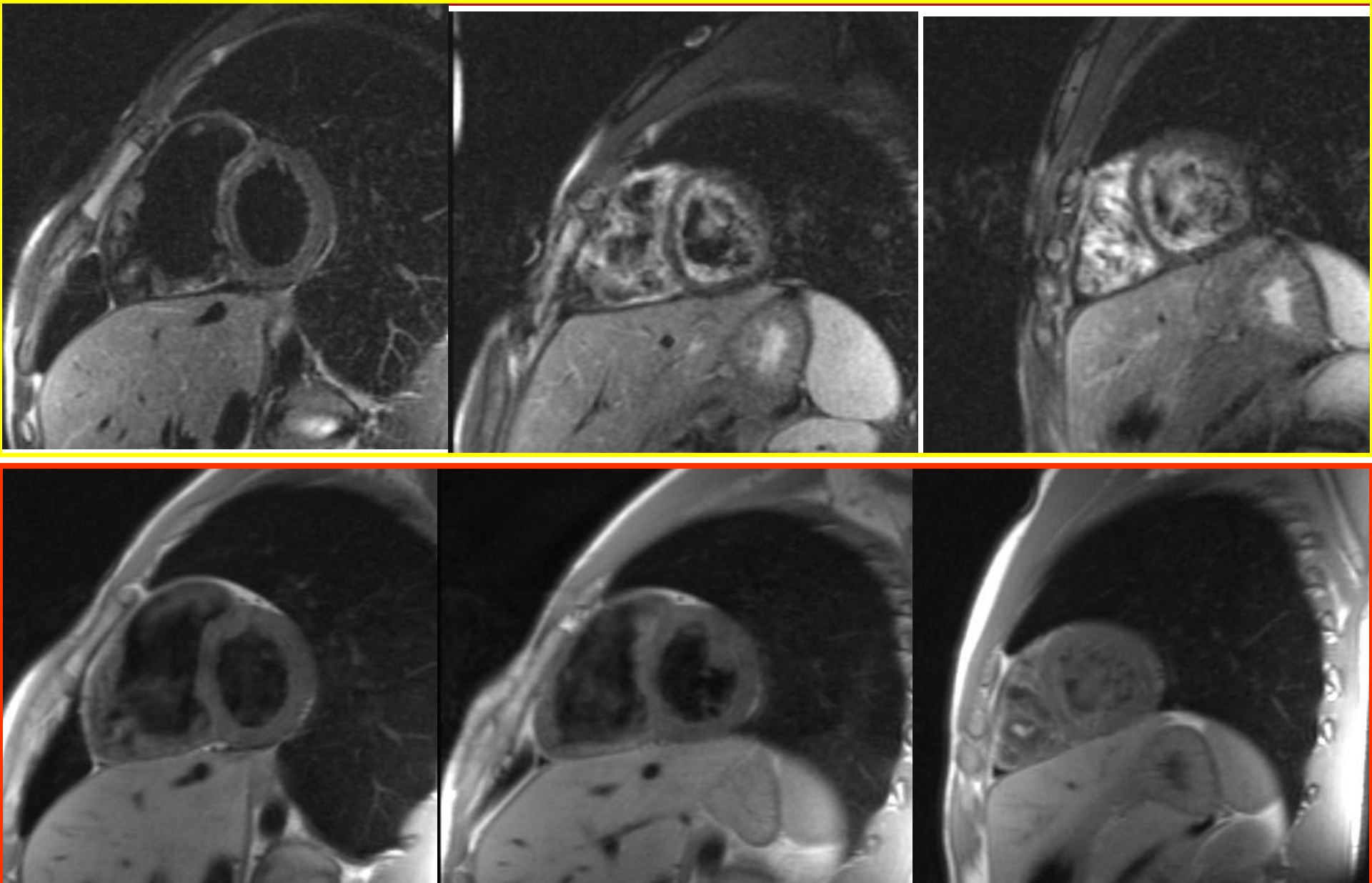

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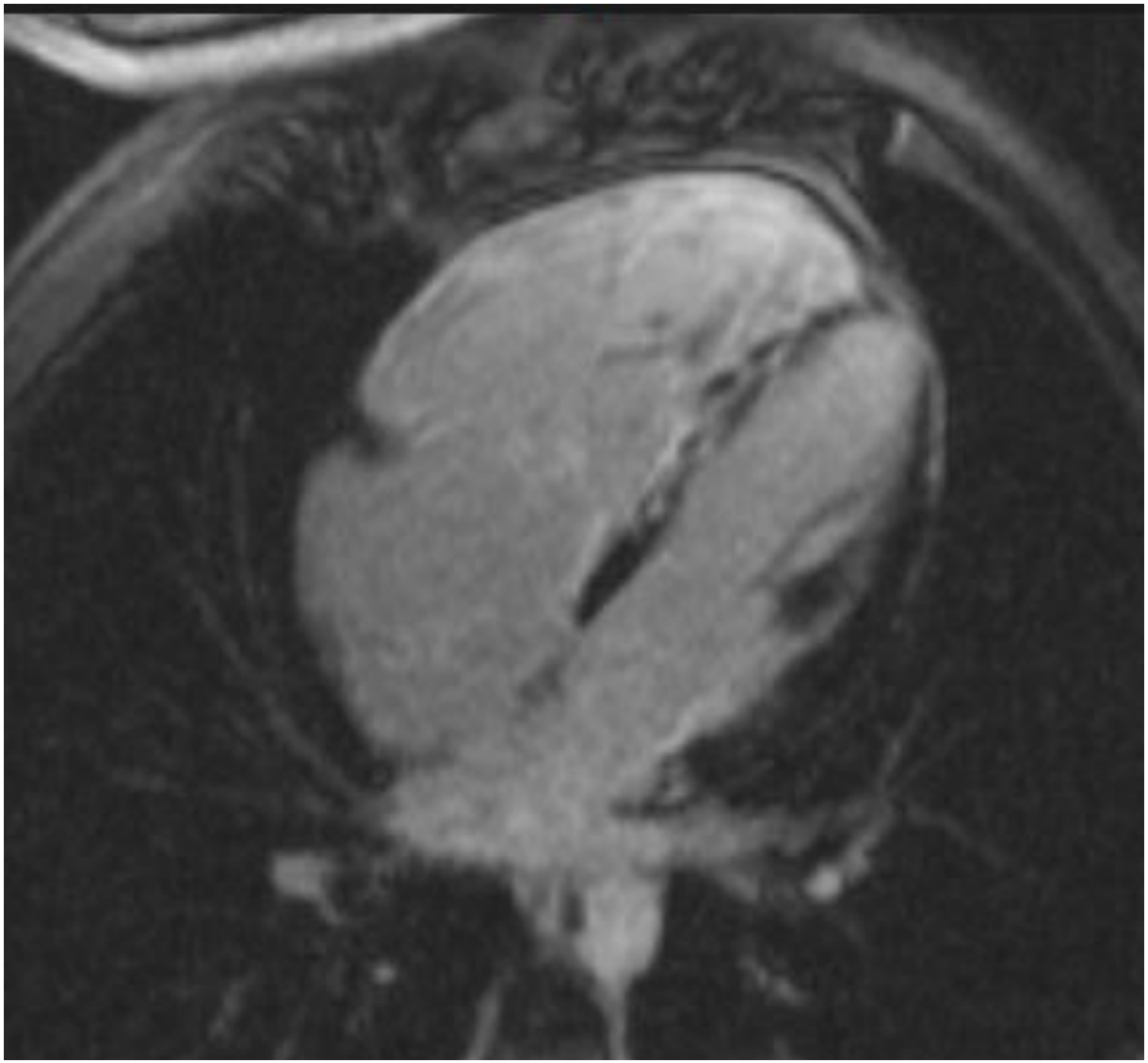

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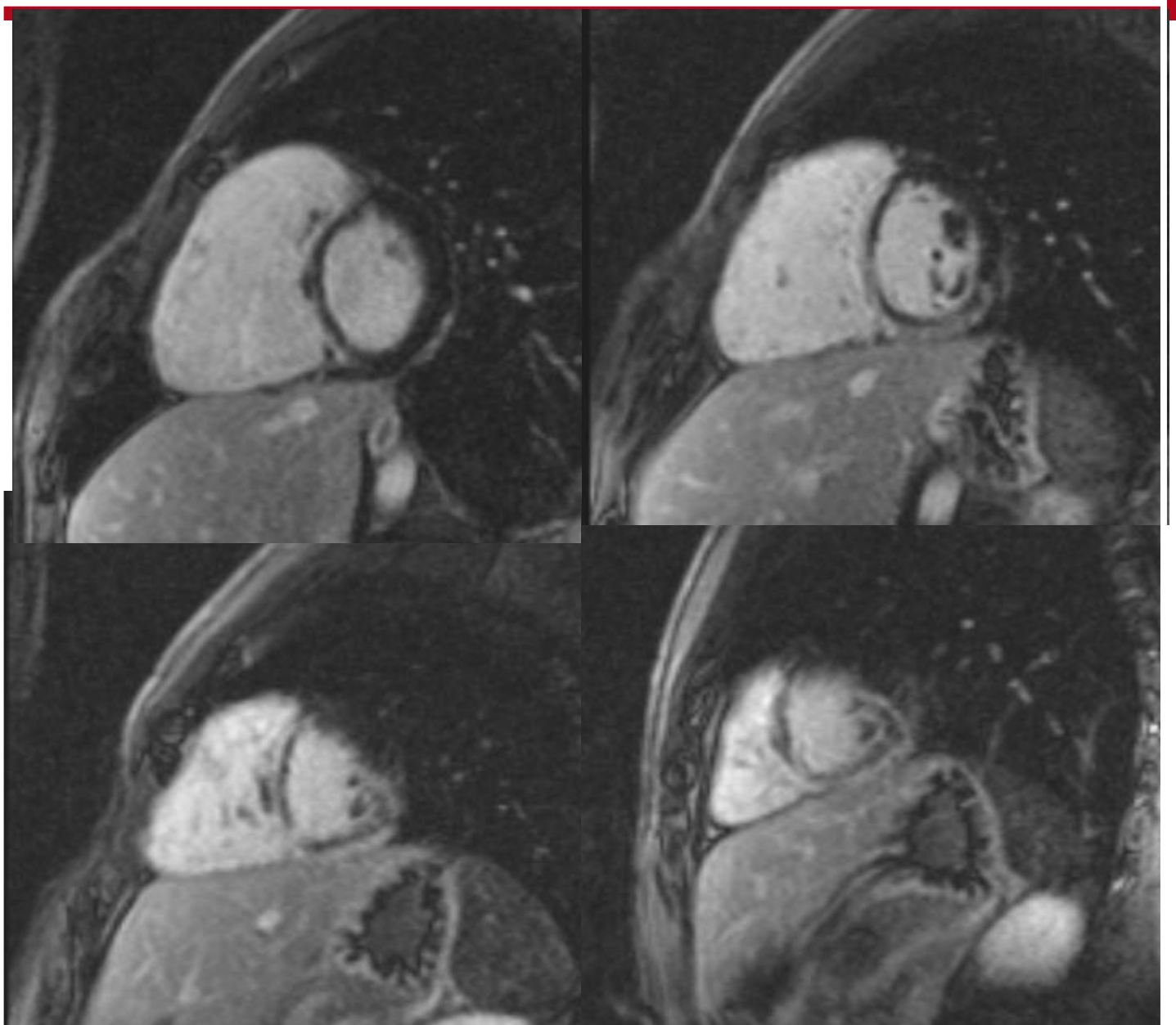

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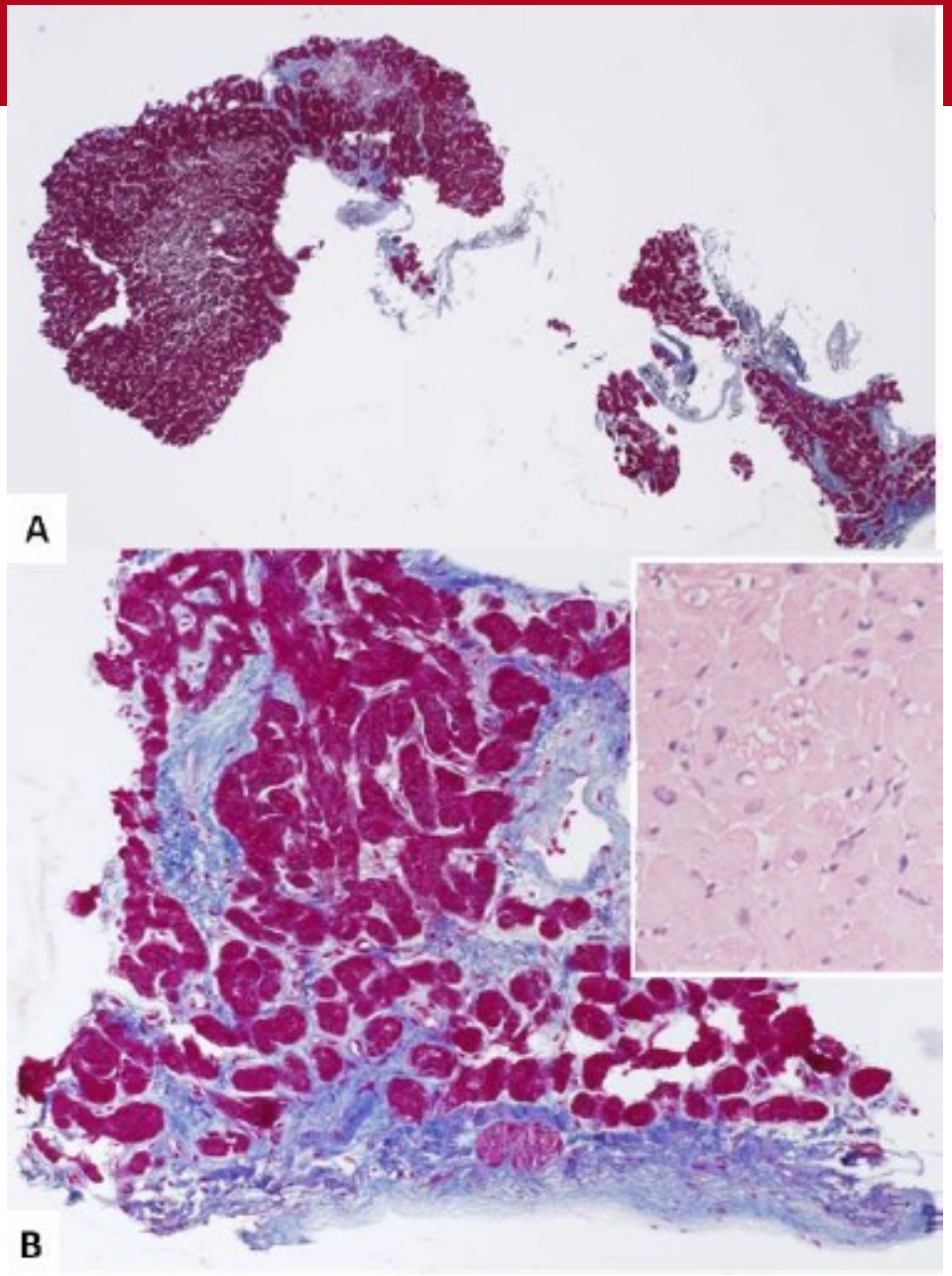


CMP a fenotipo «non dilatativo e non ipertrofico»

CARDIOMIOPATIA ARITMOGENA «LEFT DOMINANT»

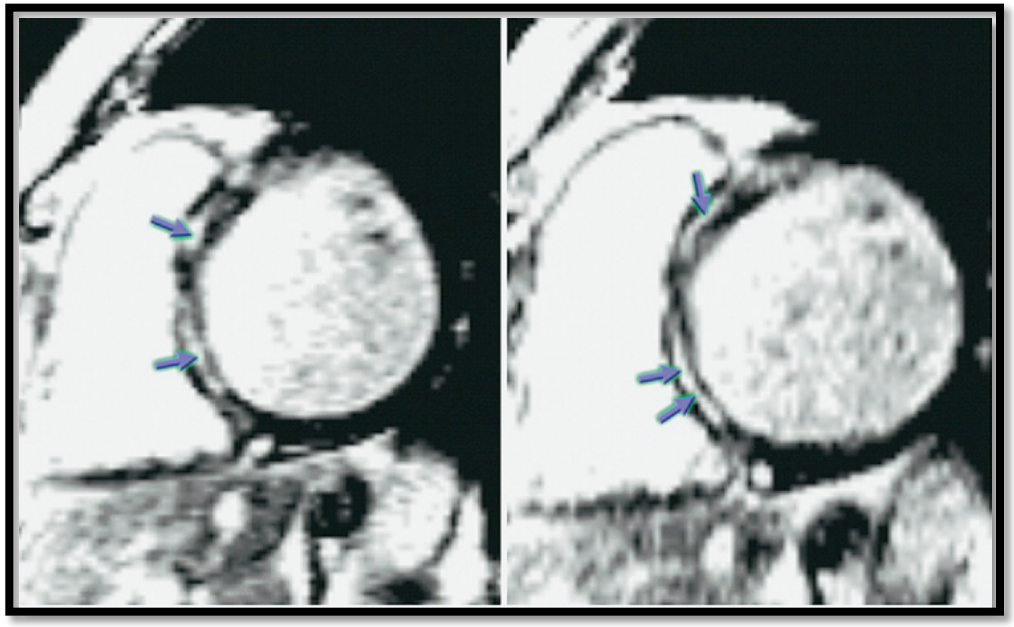
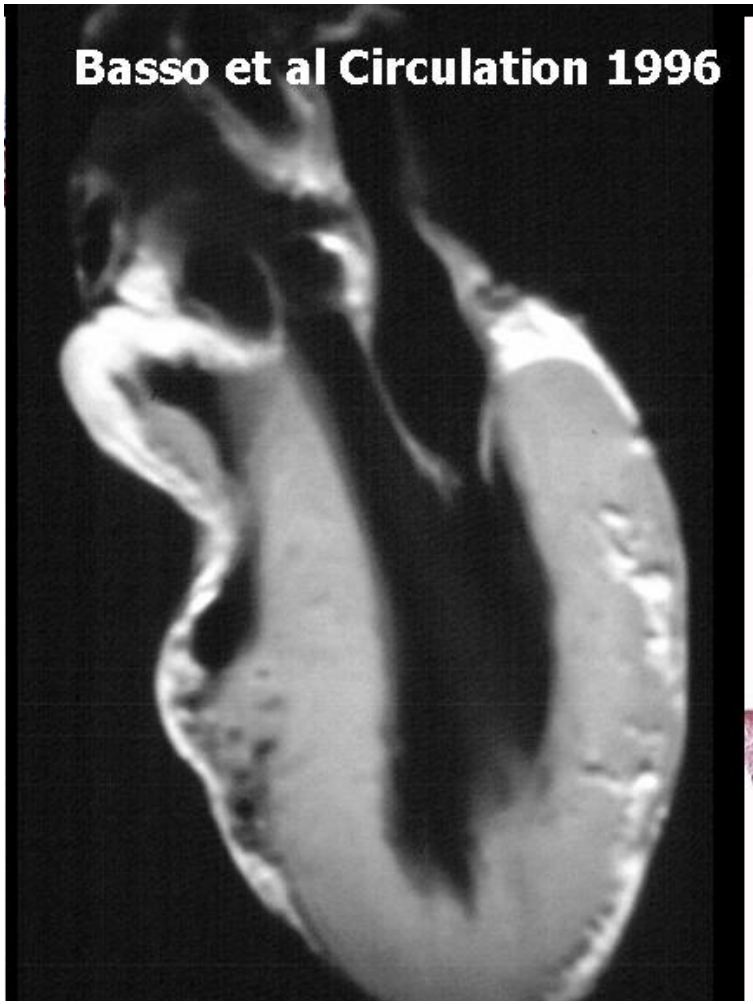


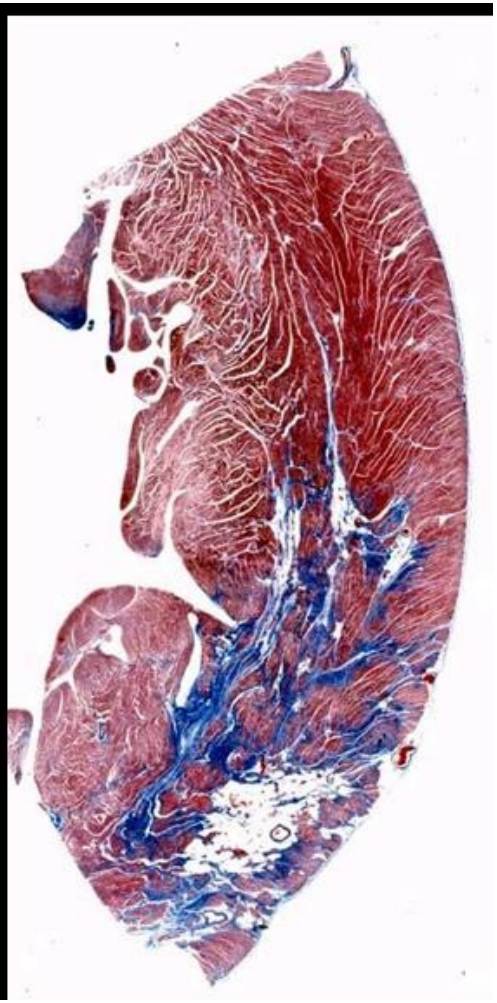
Table 6 Clinical Diagnostic Features of LDAC*

ECG	Unexplained T-wave inversion in V_5, $V_6 \pm V_4$, I, and aVL
Arrhythmia	Sustained or nonsustained ventricular tachycardia of RBBB configuration documented on ECG or Holter monitoring or during exercise testing
	Frequent ventricular extrasystoles (RBBB morphology)
Imaging	LV aneurysms
	Mild LV dilation and/or systolic impairment (with arrhythmic presentation)*
Biopsy/CMR	Myocyte loss with fibrofatty replacement on histology
	Extensive LGE of LV myocardium (with subepicardial/midmyocardial distribution)

Chowdhry et al. JACC 2008

CMP a fenotipo «non dilatativo e non ipertrofico»

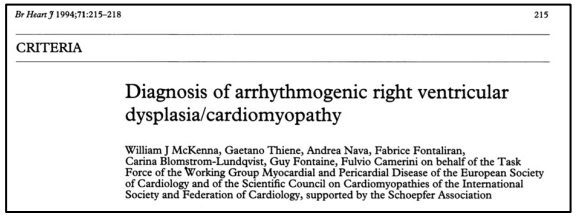




Il VS era coinvolto in più del 50% dei casi.

Nuovo approccio alla Cardiomiopatia Aritmogena

Once upon a time……

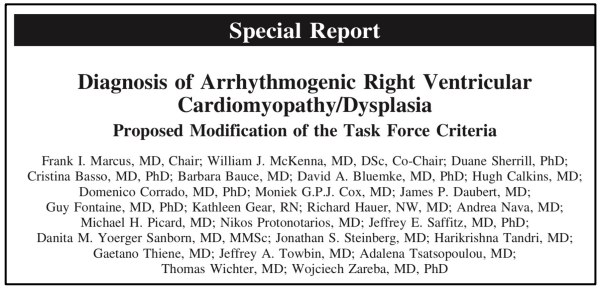


Br Heart J 1994;71:215–218 215

CRITERIA

Diagnosis of arrhythmogenic right ventricular dysplasia/cardiomyopathy

William J McKenna, Gaetano Thiene, Andrea Nava, Fabrice Fontaliran, Carina Blomstrom-Lundqvist, Guy Fontaine, Fulvio Camerini on behalf of the Task Force of the Working Group Myocardial and Pericardial Disease of the European Society of Cardiology and of the Scientific Council on Cardiomyopathies of the International Society and Federation of Cardiology, supported by the Schoepfer Association



Special Report

Diagnosis of Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia

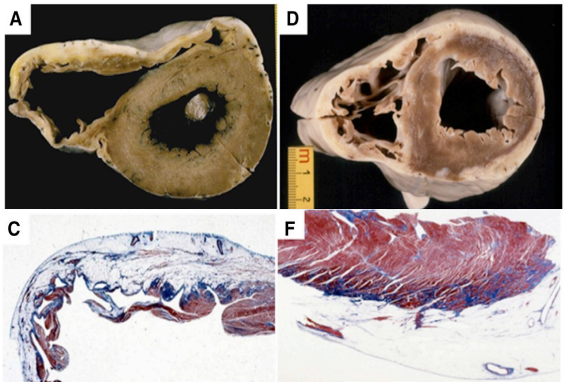
Proposed Modification of the Task Force Criteria

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

199 201

Nuovo approccio alla Cardiomiopatia Aritmogena

Once upon a time……



Corrado D. Circulation Research. 2017 (784–802)

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1994 **2010** **2023**

CMR & LGE

Diagnosis of arrhythmogenic cardiomyopathy: The Padua criteria

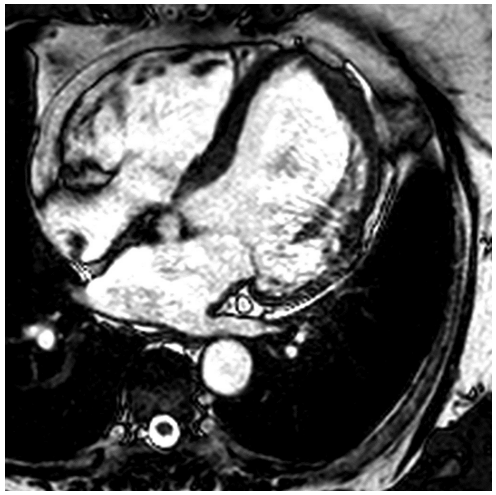
Domenico Corrado [a,*], Martina Perazzolo Marra [a], Alessandro Zorzi [a], Giorgia Beffagna [a], Alberto Cipriani [a], Manuel De Lazzari [a], Federico Migliore [a], Kalliopi Pilichou [a], Alessandra Rampazzo [b], Ilaria Rigato [a], Stefania Rizzo [a], Gaetano Thiene [a], Aris Anastasakis [c], Angeliki 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]

Nuovo approccio alla Cardiomiopatia Aritmogena

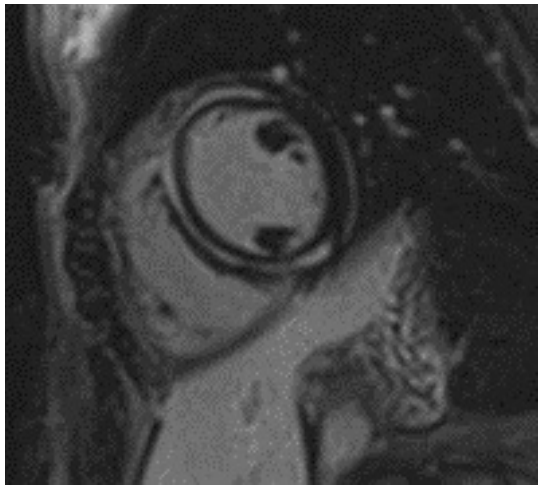
ESC European Society of Cardiology
European Heart Journal (2019) **0**, 1–16
doi:10.1093/eurheartj/ehz669

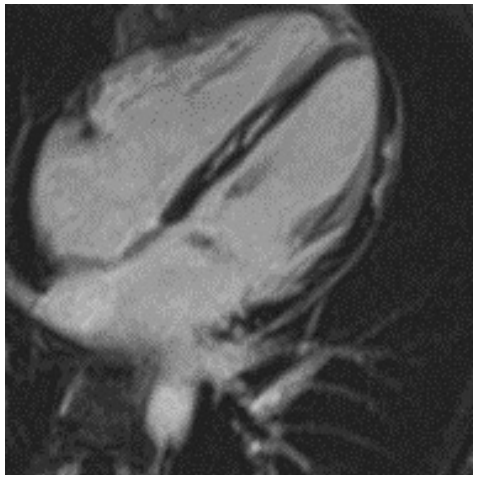
CURRENT OPINION
Heart failure/cardiomyopathy

Arrhythmogenic right ventricular cardiomyopathy: evaluation of the current diagnostic criteria and differential diagnosis



ARVC= echo



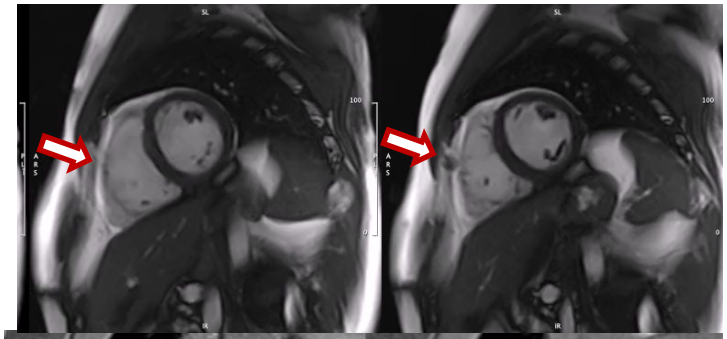


AC= CMR

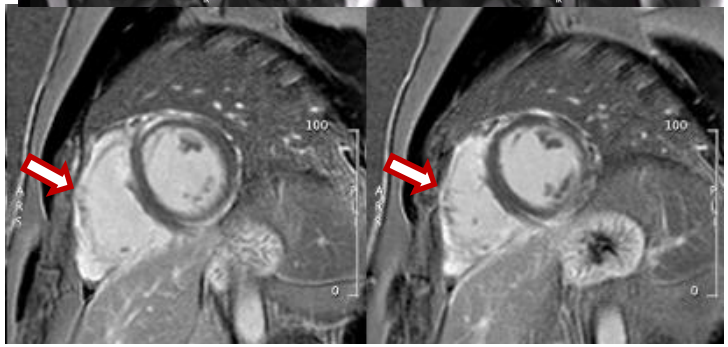
Nuovo approccio alla Cardiomiopatia Aritmogena

CONCORDANZA CINE-LGE

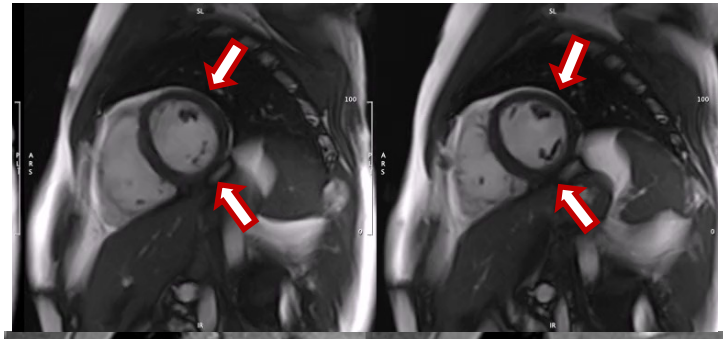
CASO 2

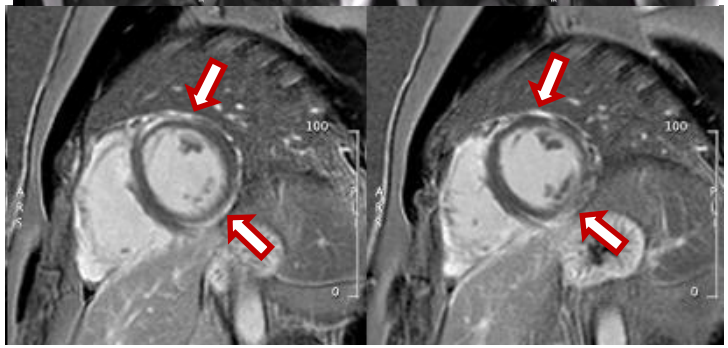


Cine asse corto.



Asse corto post-contrasto.





SI PER VDX

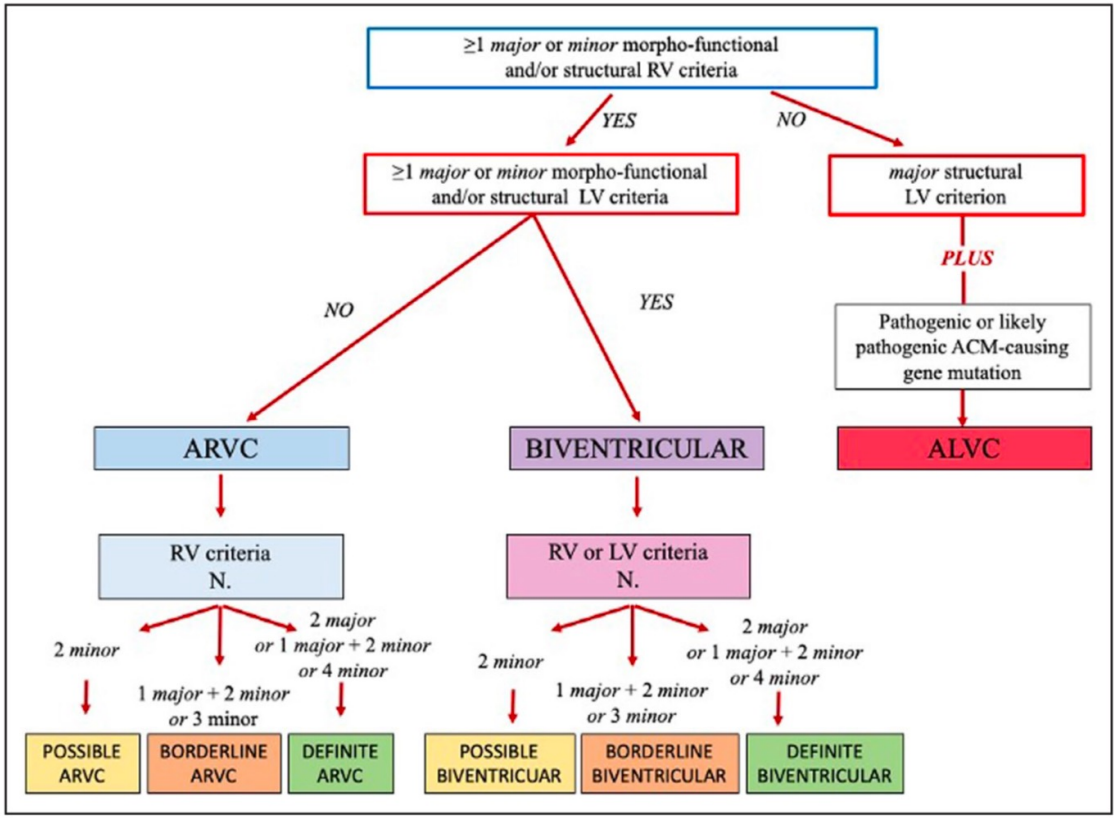
NO PER VSN

Nuovo approccio alla Cardiomiopatia Aritmogena

CONTEMPORARY REVIEW

Evolving Diagnostic Criteria for Arrhythmogenic Cardiomyopathy

Diagnostic flow-chart for ACM phenotypic variants



Corrado D et al. J Am Heart Assoc. 2021

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)

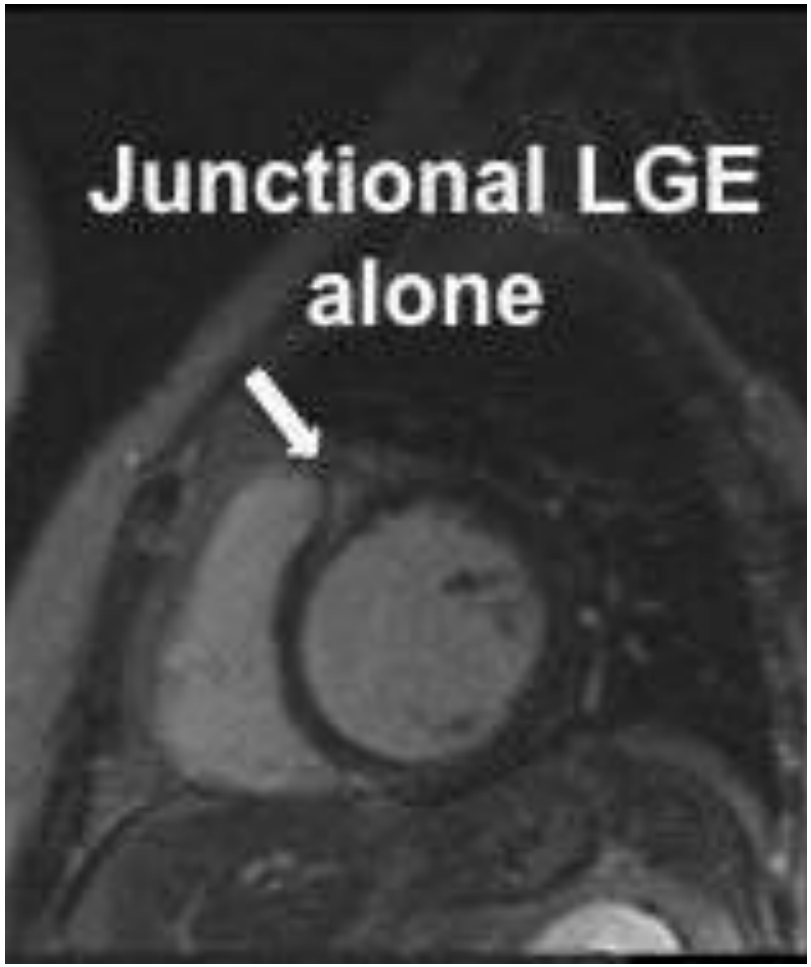
…nella cardiomiopatia aritmogena

ARVC is characterized by a predominant RV involvement. The 2010 revised international diagnostic task force criteria are based on a multiparametric strategy[116] (*Figure 21*). Tissue characterization by CMR was not included. However, RV fatty infiltration and LV LGE are frequently observed (in 29–53% and 35.5–45% of probands, respectively) and may be present before patients meet major task force imaging criteria.[676–678] Both wall motion alterations and pre-/post-contrast signal abnormalities have been suggested to enhance the diagnostic accuracy of CMR for ARVC.[679] The identification of biventricular and left-dominant involvement in ARVC patients[680,681] has recently led to the proposed term 'arrhythmogenic cardiomyopathy'.[682]

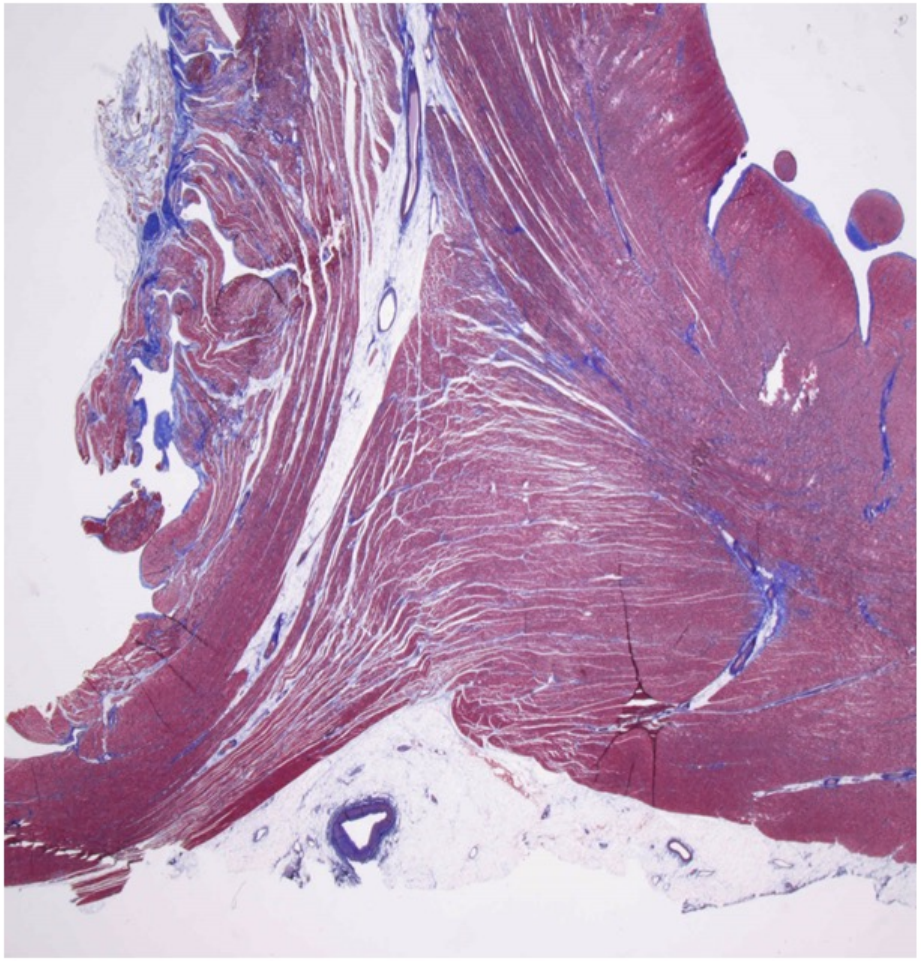
Recommendations	Class[a]	Level[b]
Diagnostic evaluation and general recommendations		
In patients with suspected ARVC, CMR is recommended.[676–678]	I	B

Ma allora se è LGE, che sia vero!!!

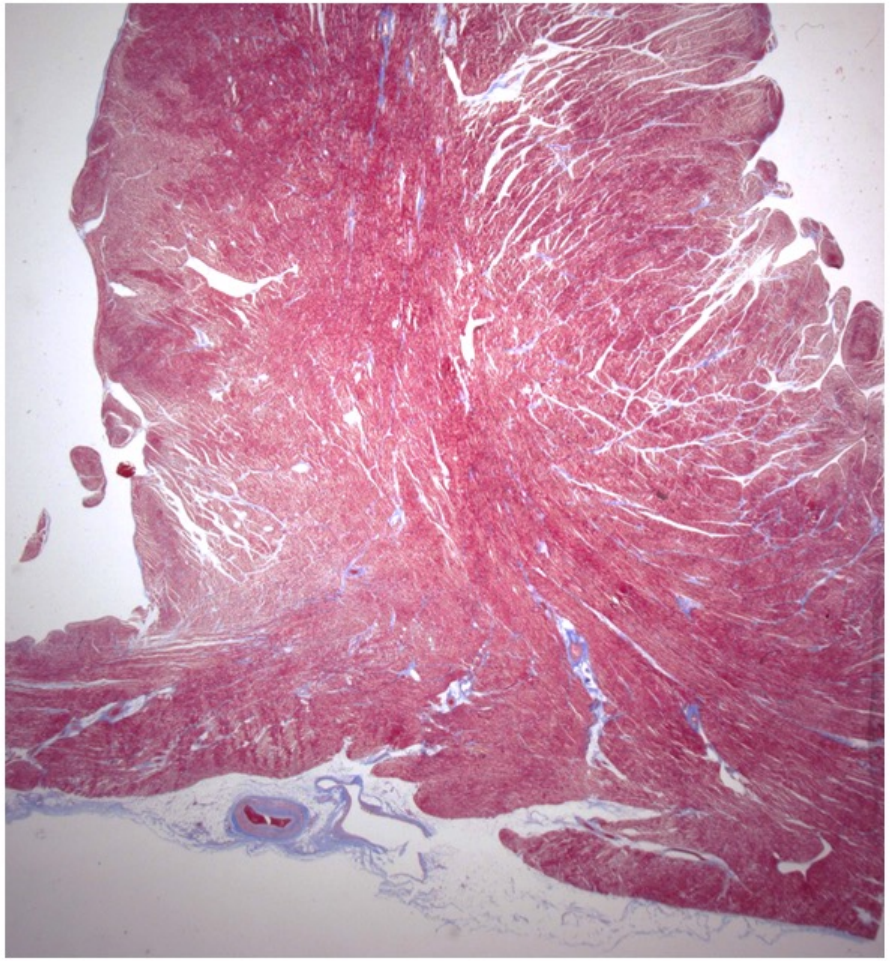
- Interpretazione di LGE “giunzionale”



Tips & Tricks
CINE IMAGES



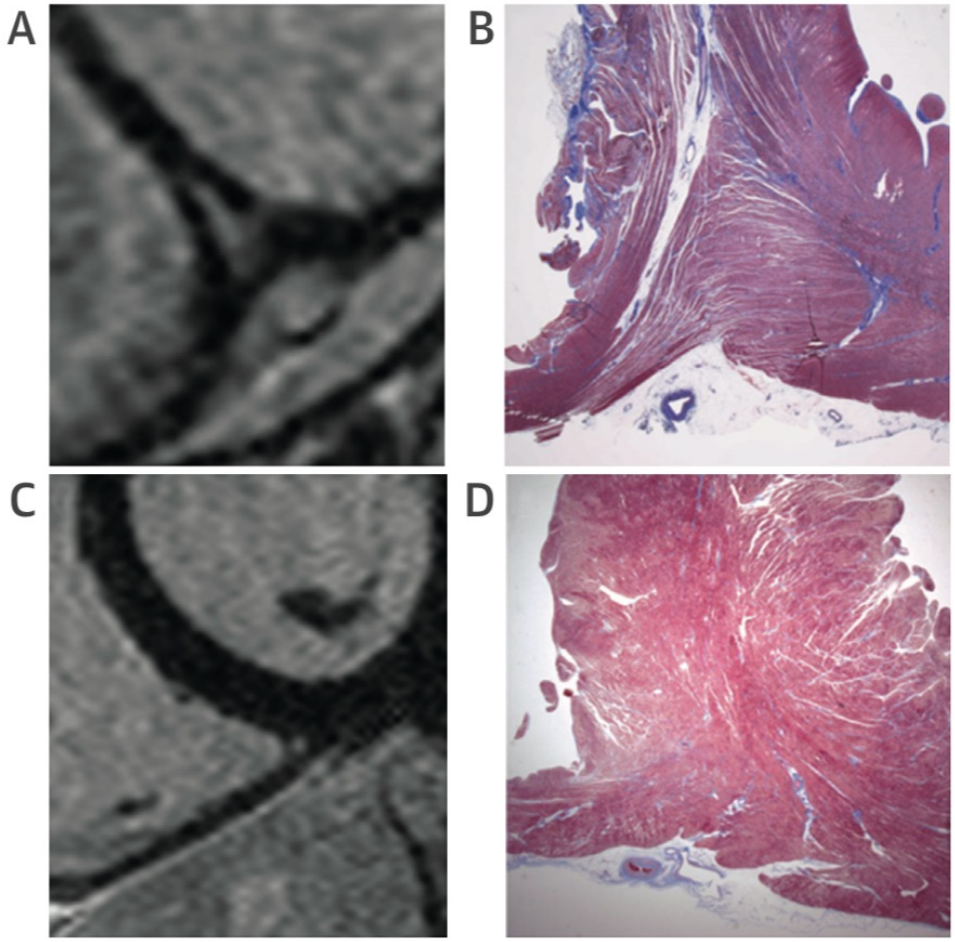
Giunzione di un paziente con DCM

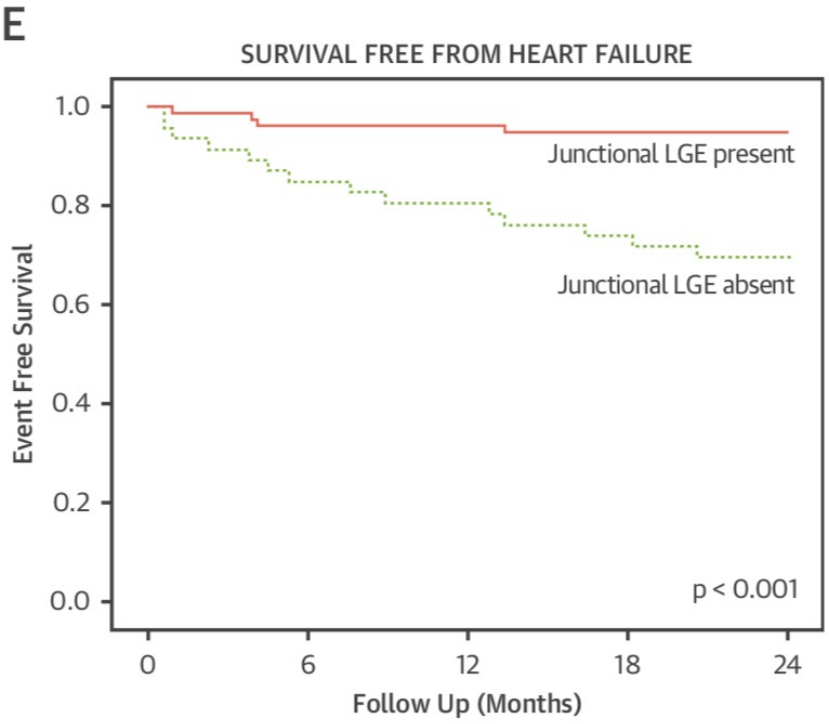


Giunzione di un soggetto normale

Right Ventricular Junctional Late Gadolinium Enhancement Correlates With Outcomes in Pulmonary Hypertension

FIGURE 1 Histological Basis of Junctional LGE in Nonischemic Dilated Cardiomyopathy





De Lazzari M et al JACC: CARDIOVASCULAR IMAGING 2019

Tips & Tricks
LGE

RESEARCH Open Access

What is the mid-wall linear high intensity "lesion" on cardiovascular magnetic resonance late gadolinium enhancement?

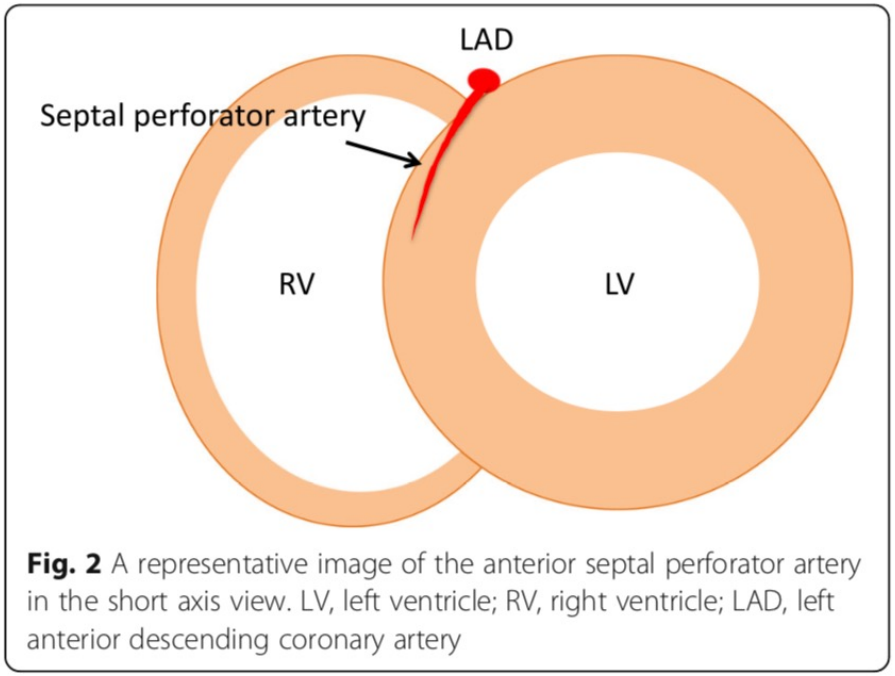
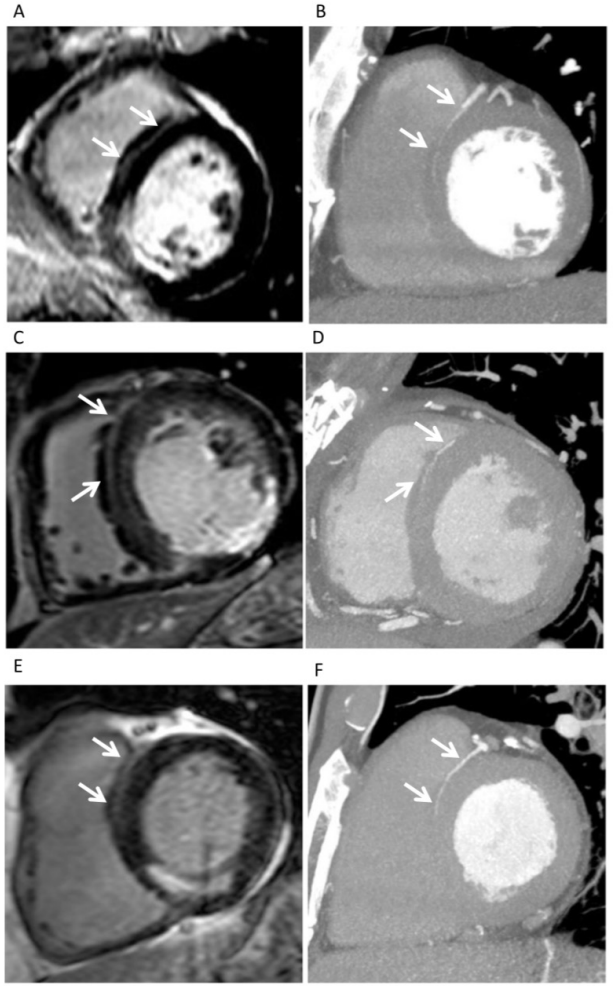


Fig. 2 A representative image of the anterior septal perforator artery in the short axis view. LV, left ventricle; RV, right ventricle; LAD, left anterior descending coronary artery

Nakamura et al. Journal of Cardiovascular Magnetic Resonance (2020) 22:66

➢ LGE settale



RESEARCH Open Access

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Check for updates

➢ LGE settale

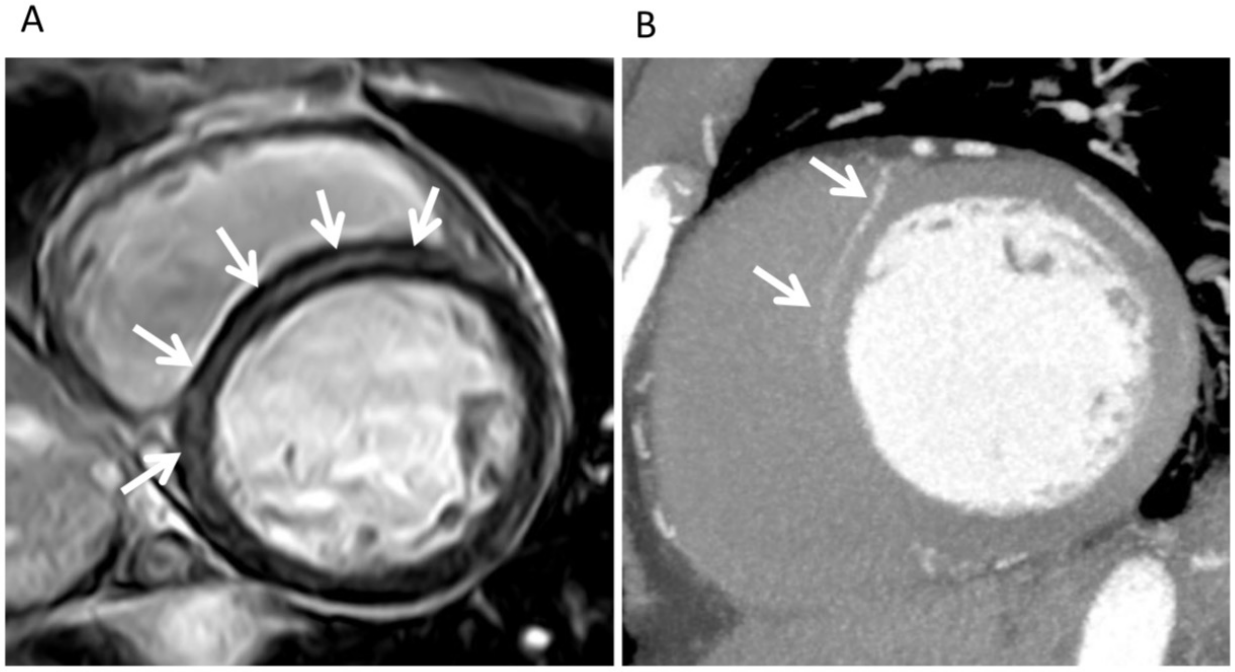
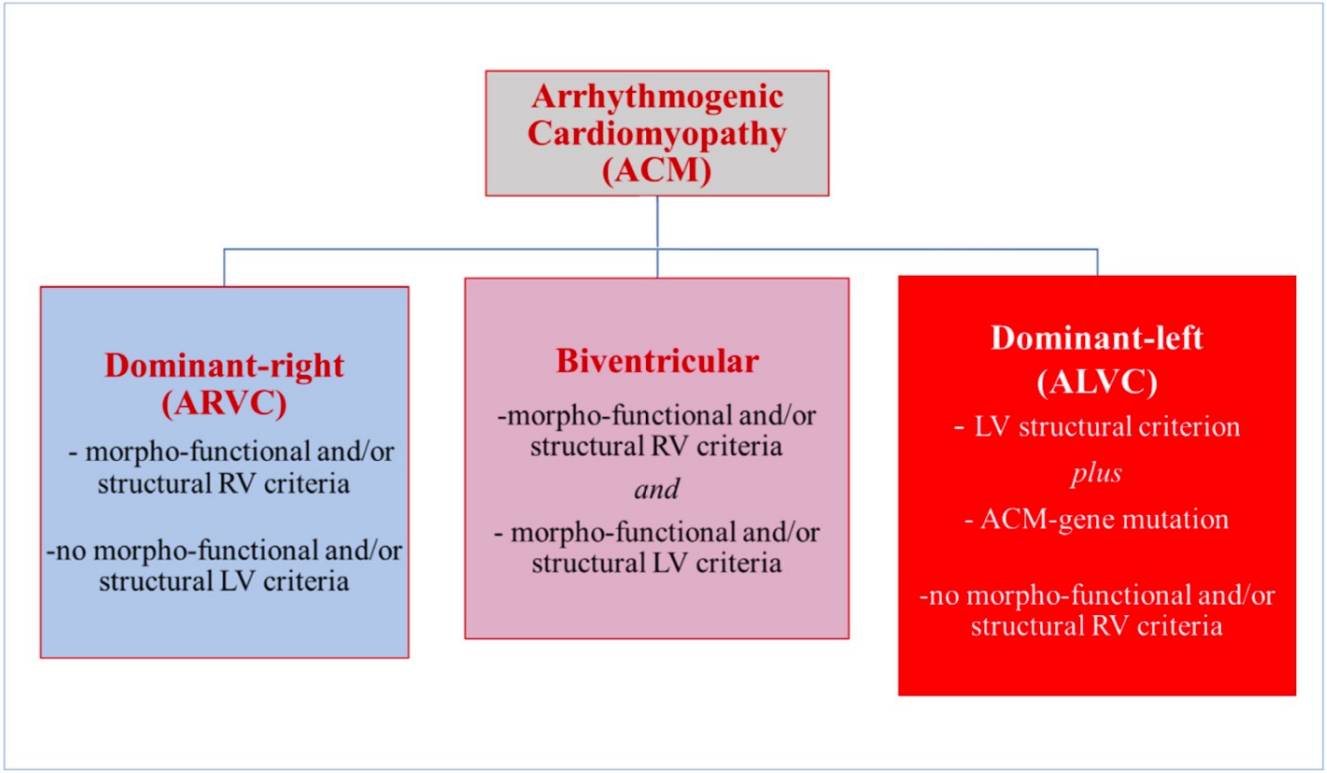


Fig. 6 Forty-six-year-old, male, dilated cardiomyopathy. Typical mid-wall fibrosis is seen in the mid-layer of the basal septum from the anterior to the inferior side (**a**; arrow). The LGE pattern is distinctly different from that of the anterior septal perforator artery observed using CorCTA on another day (**b**; arrow)

Nakamura et al. Journal of Cardiovascular Magnetic Resonance (2020) 22:66

Dalla teoria alla pratica…

- La diagnosi differenziale della forma ARVC
- La diagnosi differenziale della forma ALVC
- La diagnosi differenziale nelle forme dilatative



Corrado D et al, International Journal of Cardiology 319 (2020) 106–114

Dalla teoria alla pratica…

- La diagnosi differenziale della forma ARVC
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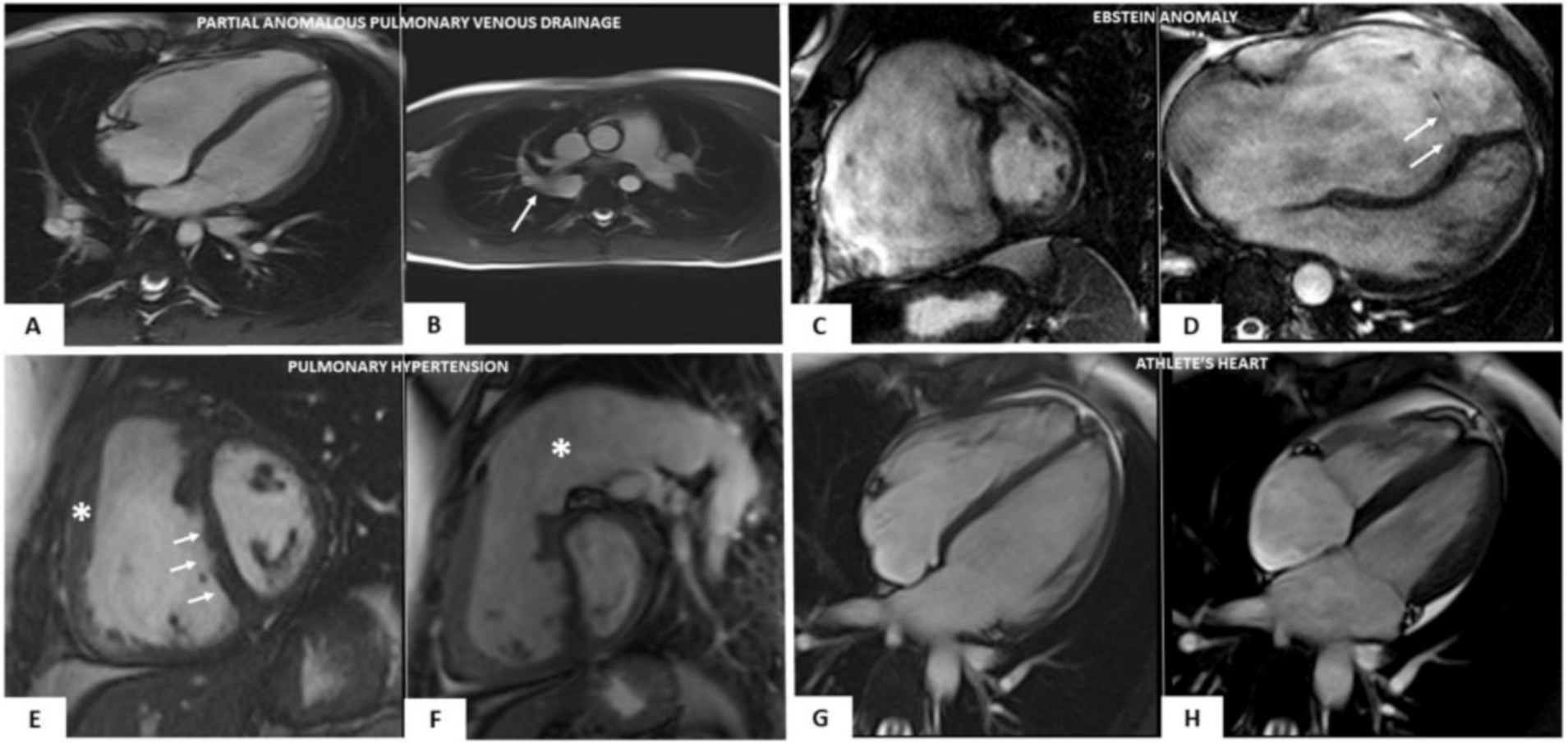
RV Phenocopies

Clinical Key Points

- Phenocopies ->mimicking RV-dominant



Dalla teoria alla pratica…

- La diagnosi differenziale della forma ARVC
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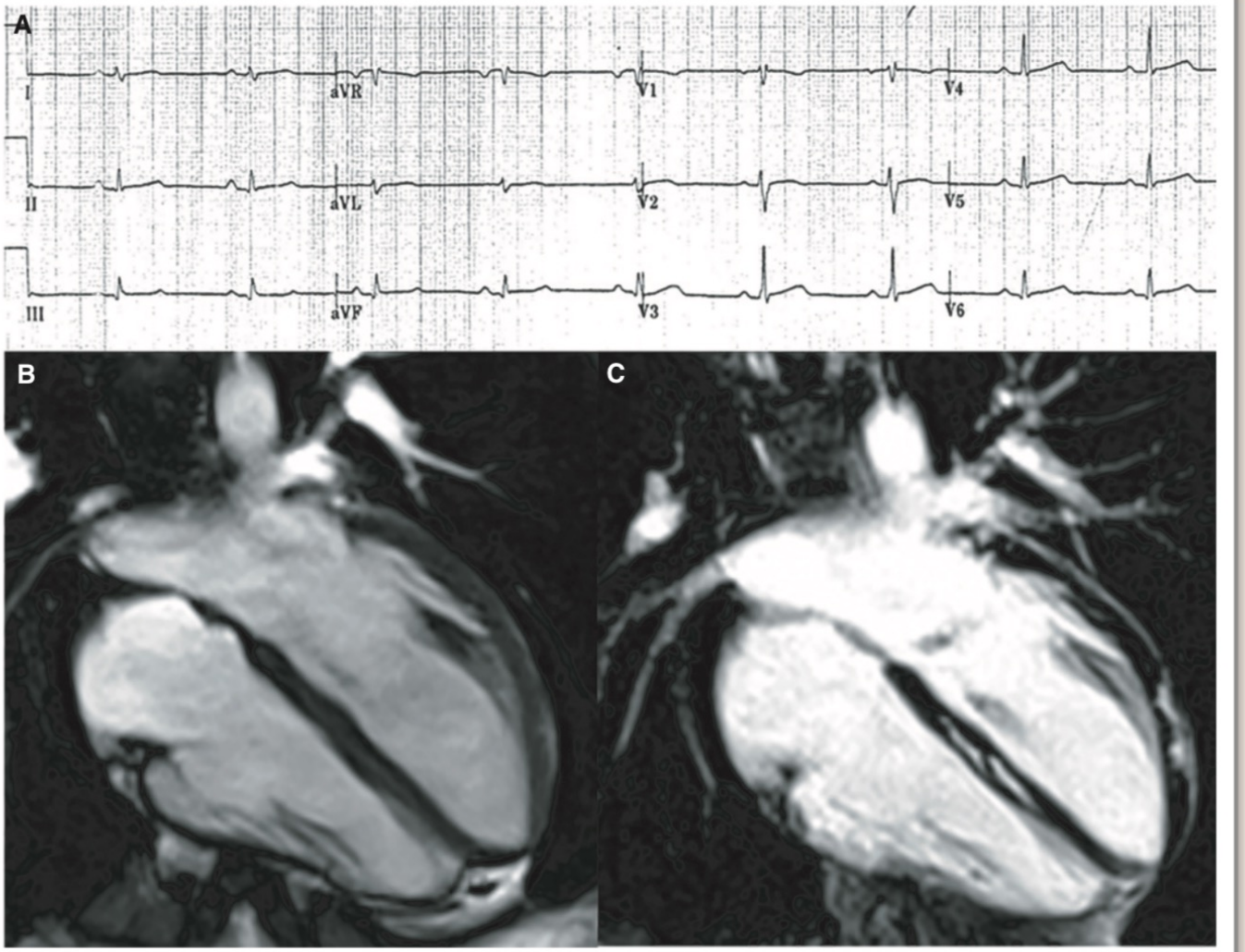
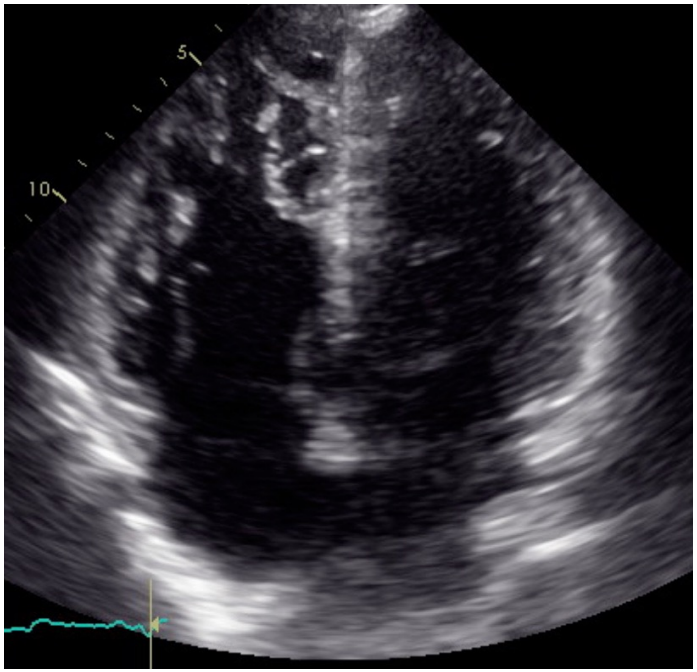
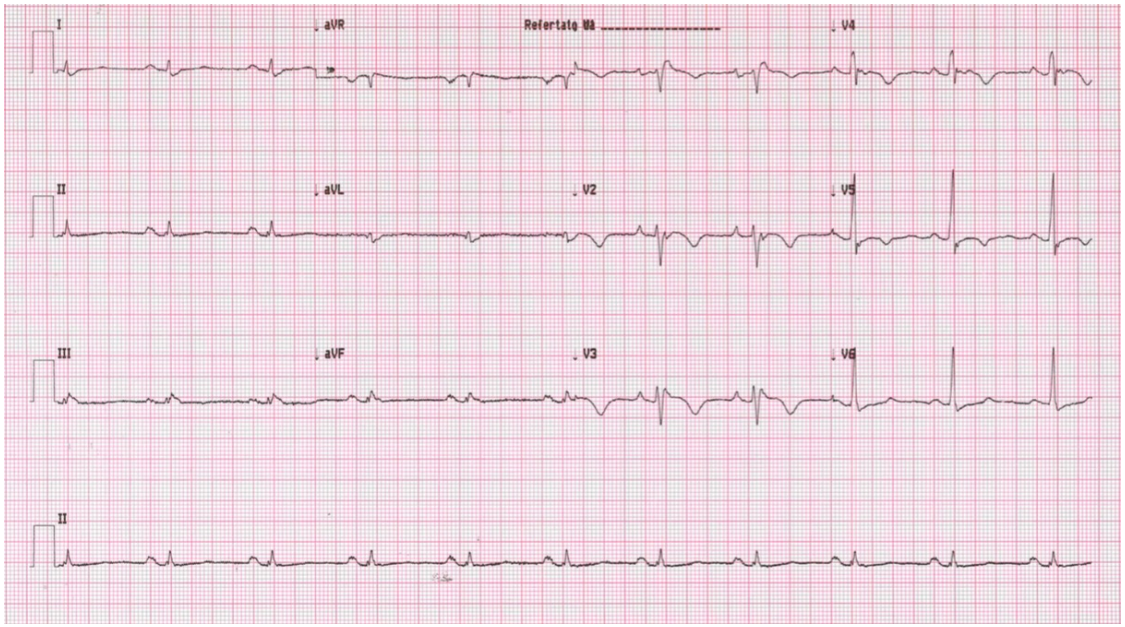
Relationship Between Electrocardiographic Findings and Cardiac Magnetic Resonance Phenotypes in Arrhythmogenic Cardiomyopathy

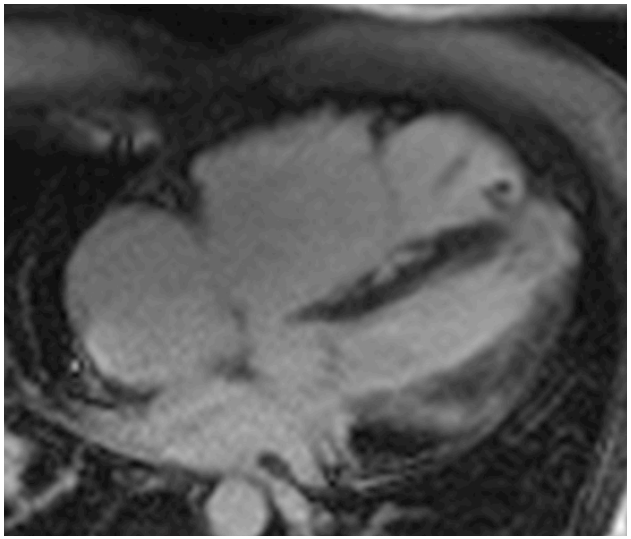


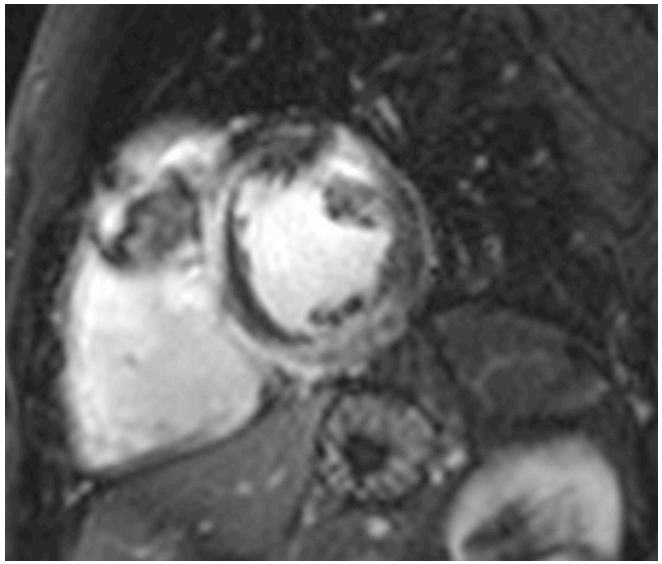
De Lazzari et al. J Am Heart Assoc. 2018

CMP a fenotipo «non dilatativo e non ipertrofico»









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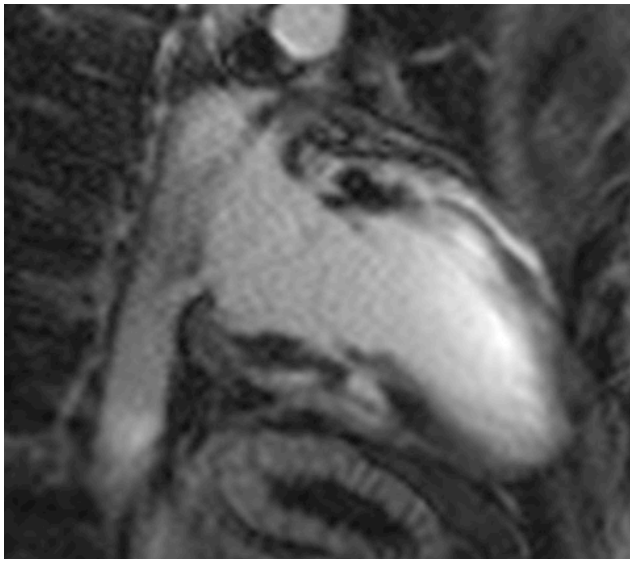
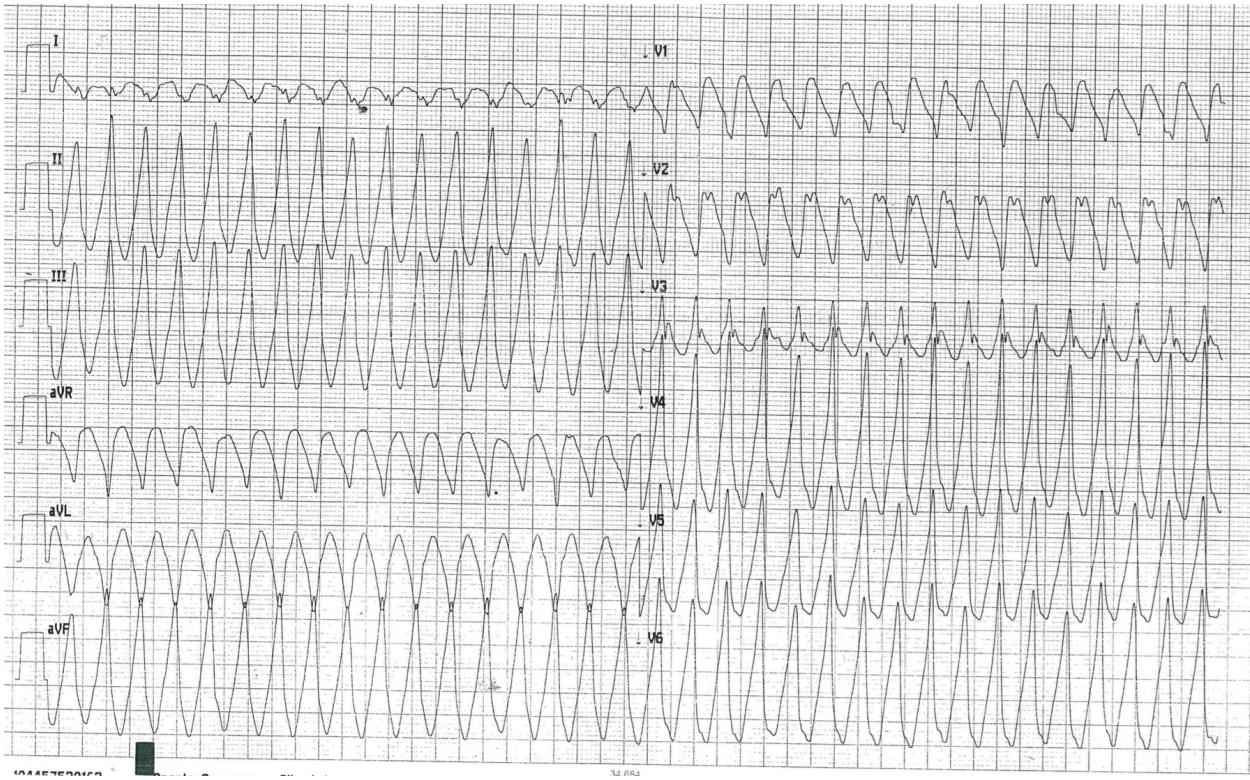


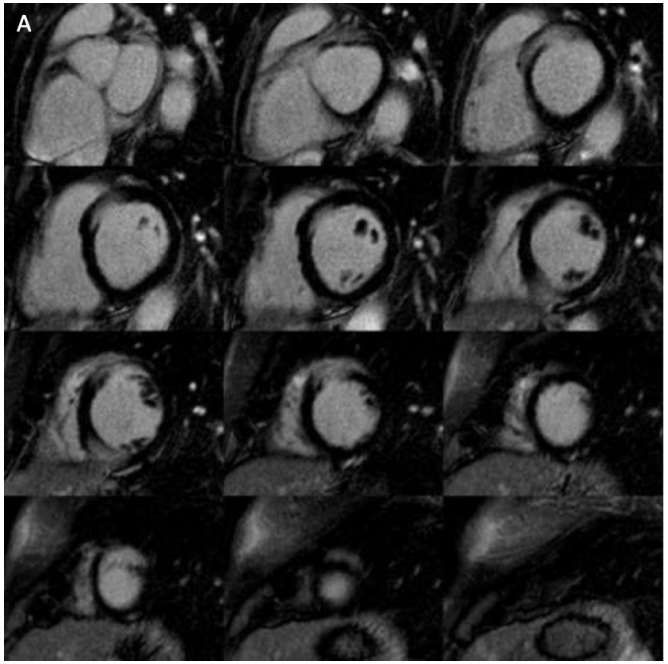
SARCOIDOSI

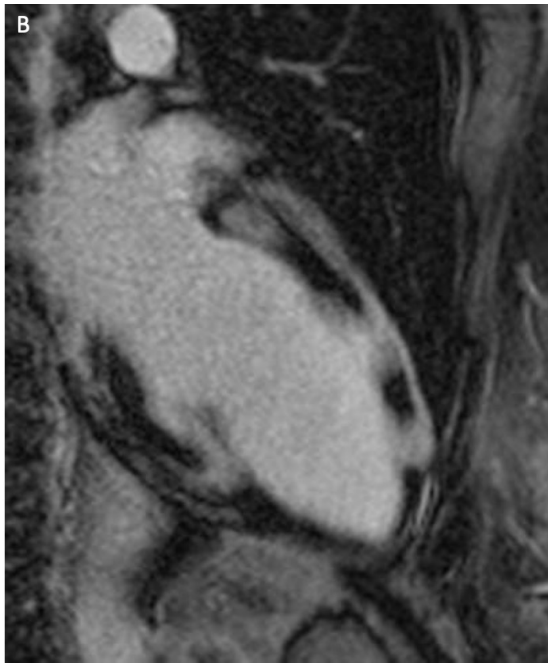


Blocco atrio-ventricolare avanzato, FA e TVNS sono comuni nella sarcoidosi.

CMP a fenotipo «non dilatativo e non ipertrofico»

SARCOIDOSI

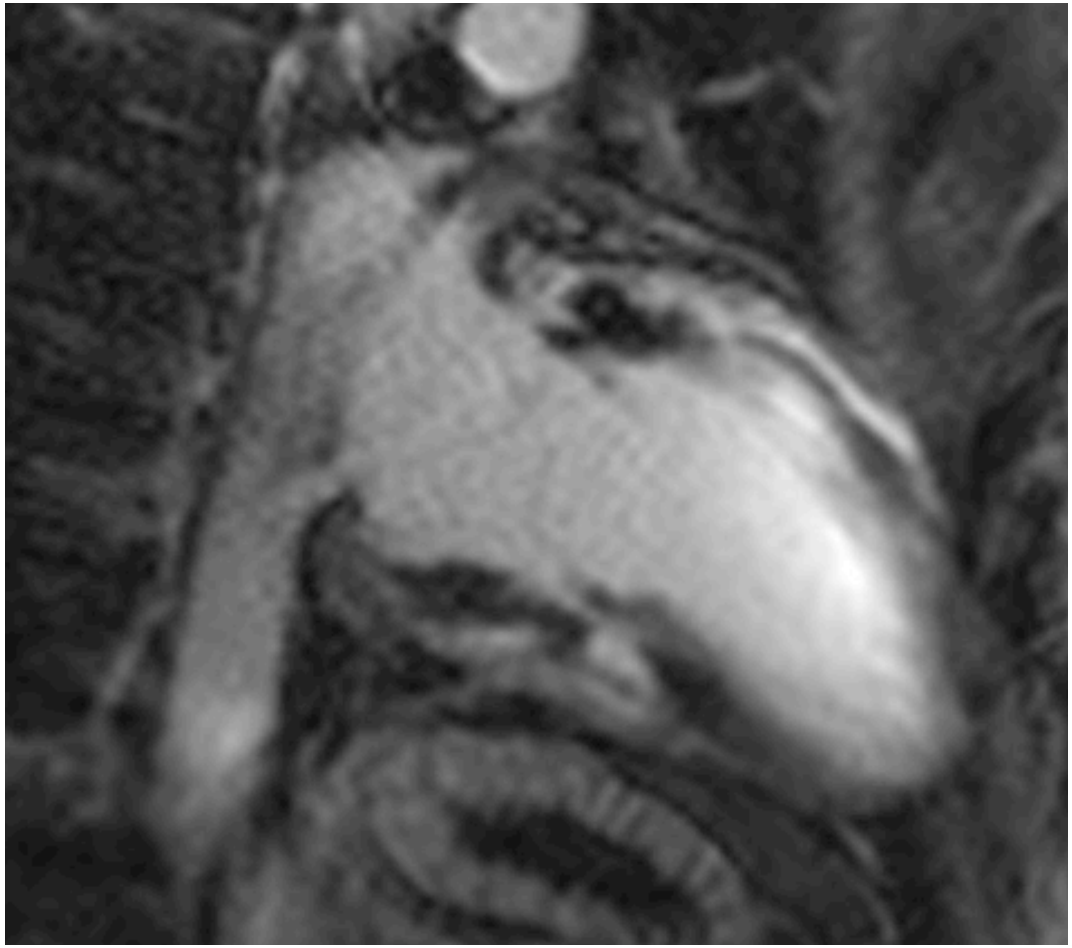


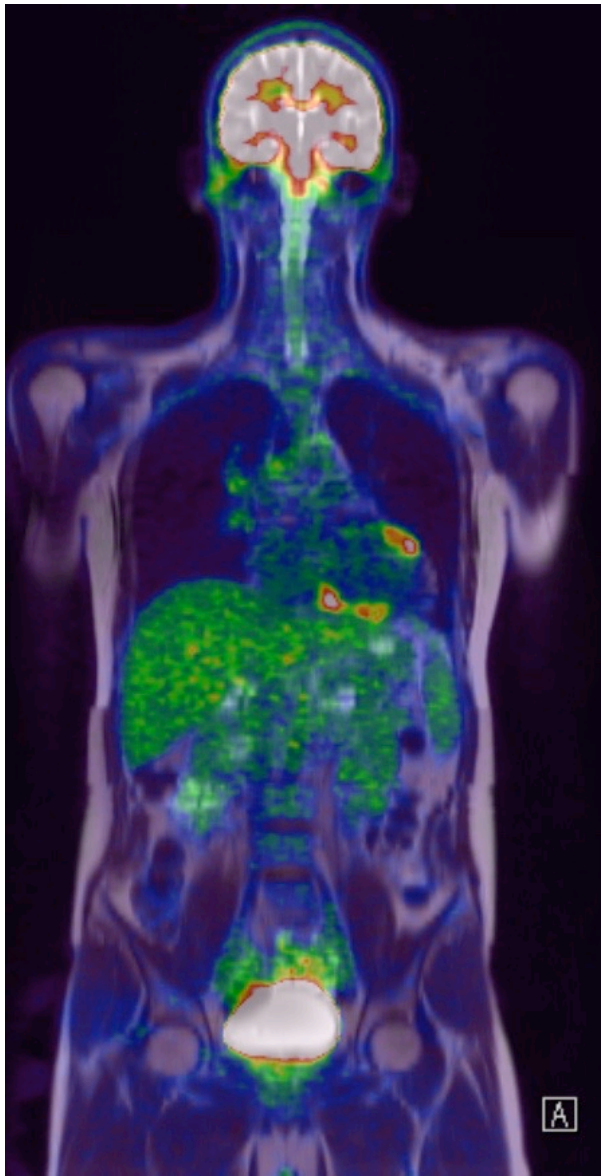


- fisiologia restrittiva del VS
- **granulomi miocardici** nelle immagini post- contrasto: **intramurali, a chiazze, prevalentemente laterale basale**. Rispondono alla terapia immunosoppressiva
- **LGE non secondo territorio di distribuzione coronarica**

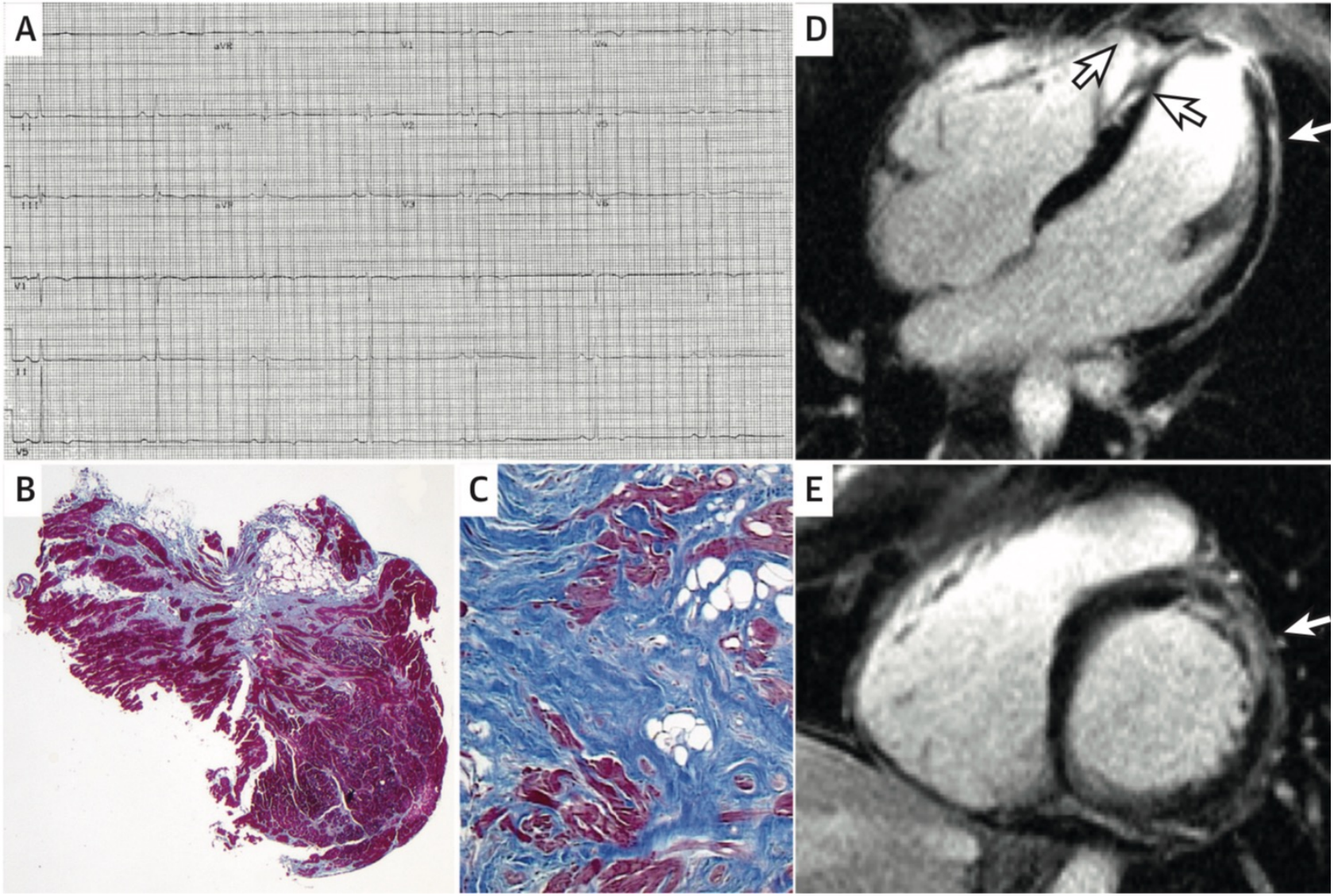
CMP a fenotipo «non dilatativo e non ipertrofico»

SARCOIDOSI



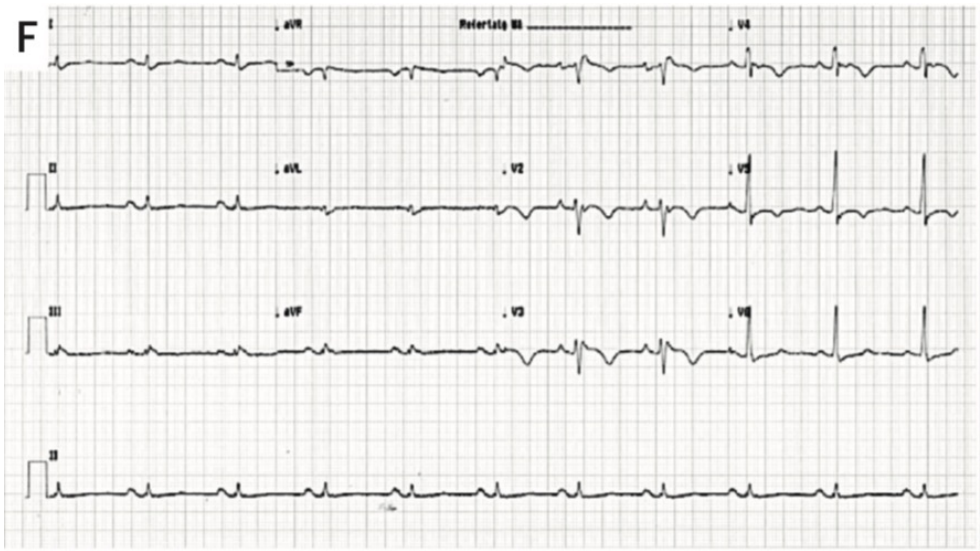


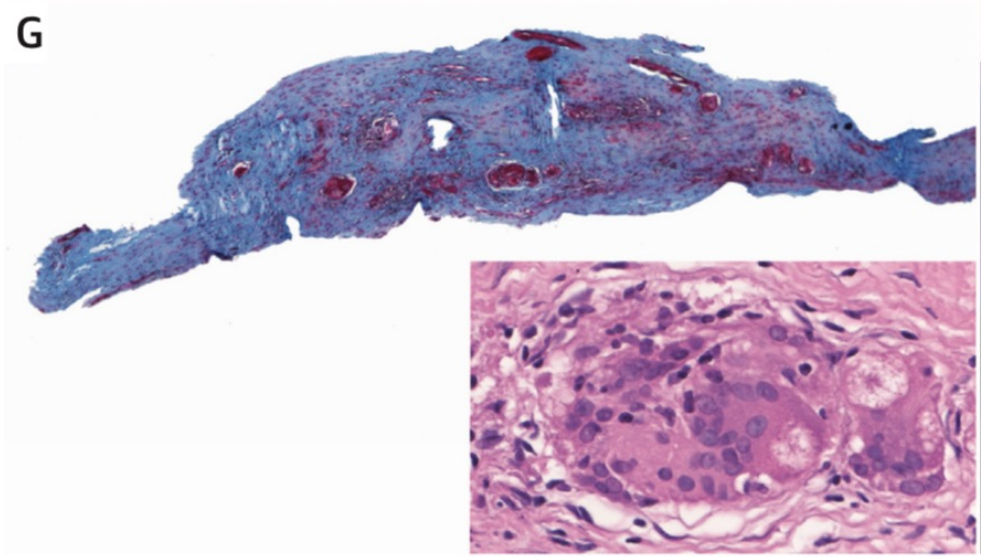
Myocardial Tissue Characterization in Arrhythmogenic Cardiomyopathy. Comparison Between Endomyocardial Biopsy and Cardiac Magnetic Resonance

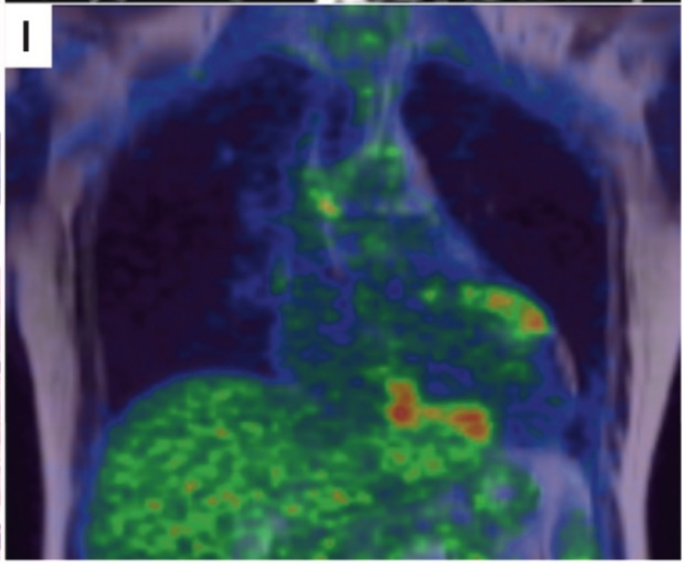


Martina Perazzolo Marra, JACC Img 2021

Myocardial Tissue Characterization in Arrhythmogenic Cardiomyopathy. Comparison Between Endomyocardial Biopsy and Cardiac Magnetic Resonance







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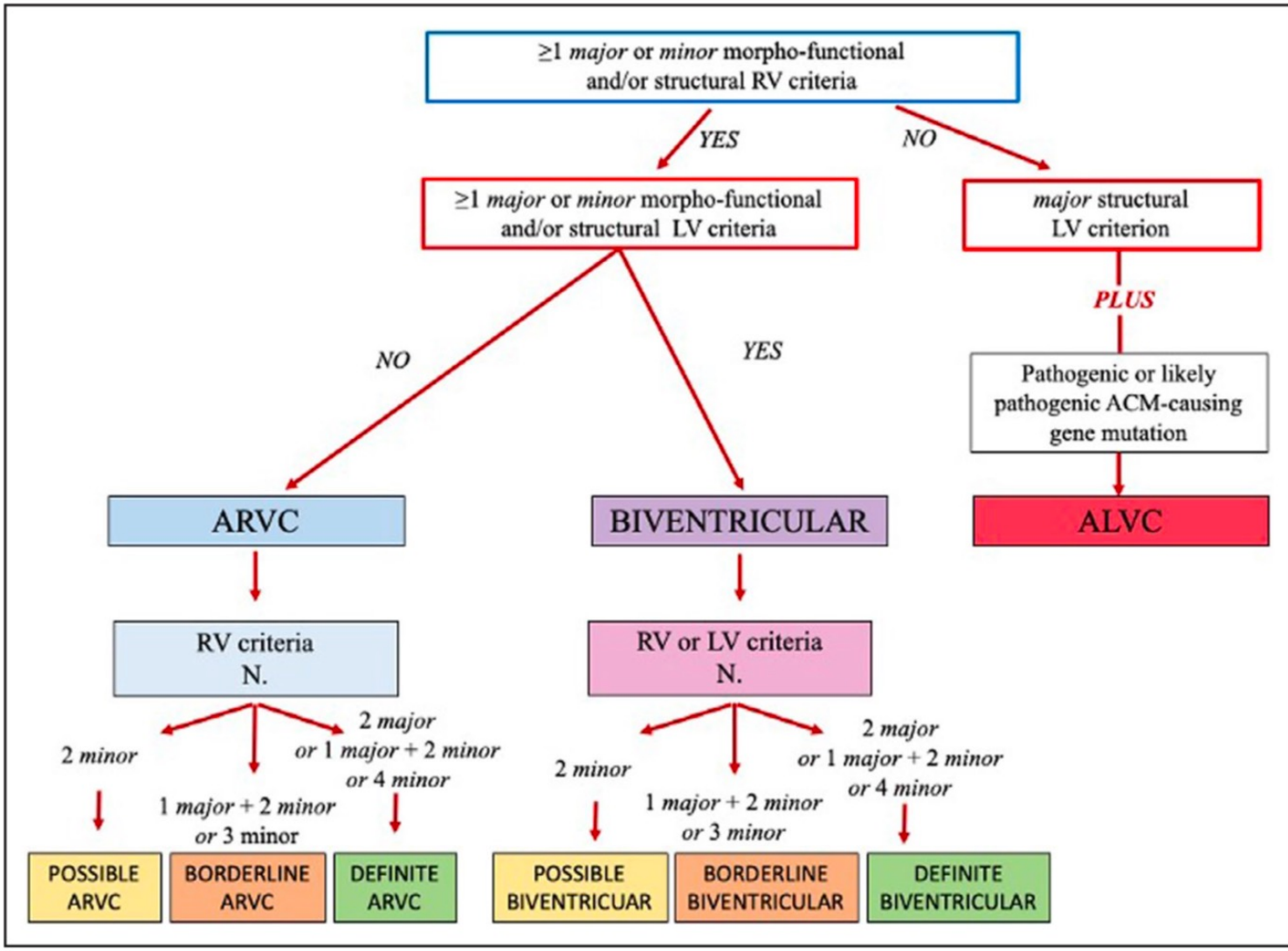
CONTEMPORARY REVIEW

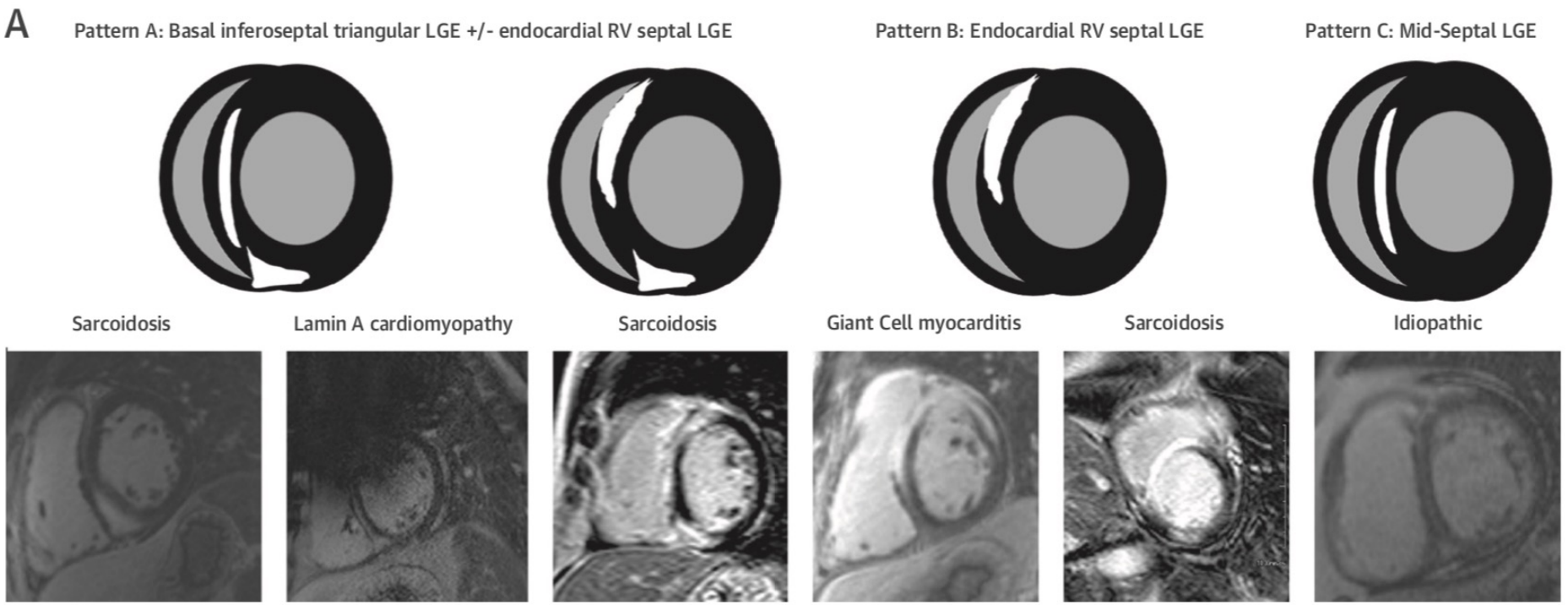
Evolving Diagnostic Criteria for Arrhythmogenic Cardiomyopathy

Corrado D et al. J Am Heart Assoc. 2021



Basal Inferoseptal Triangular Late Gadolinium Enhancement

FIGURE 1 Illustration of Septal and Inferoseptal Basal Triangular Scar Patterns in Cardiac Sarcoidosis

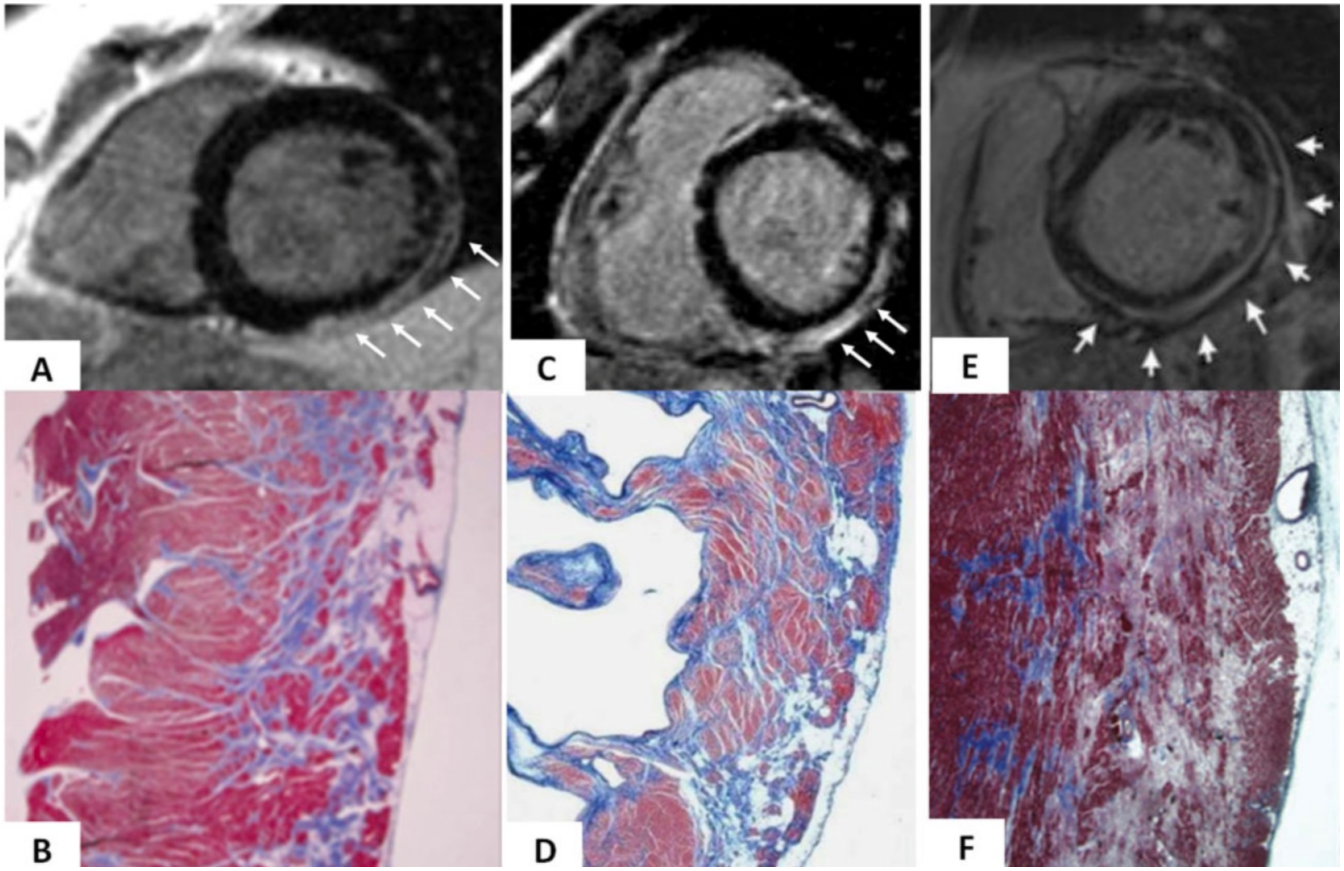


Kuo L. et al. JACC: CARDIOVASCULAR IMAGING, VOL. 12, NO. 12, 2019

Phenocopies
Neuromuscolar diseases

Clinical Key Points

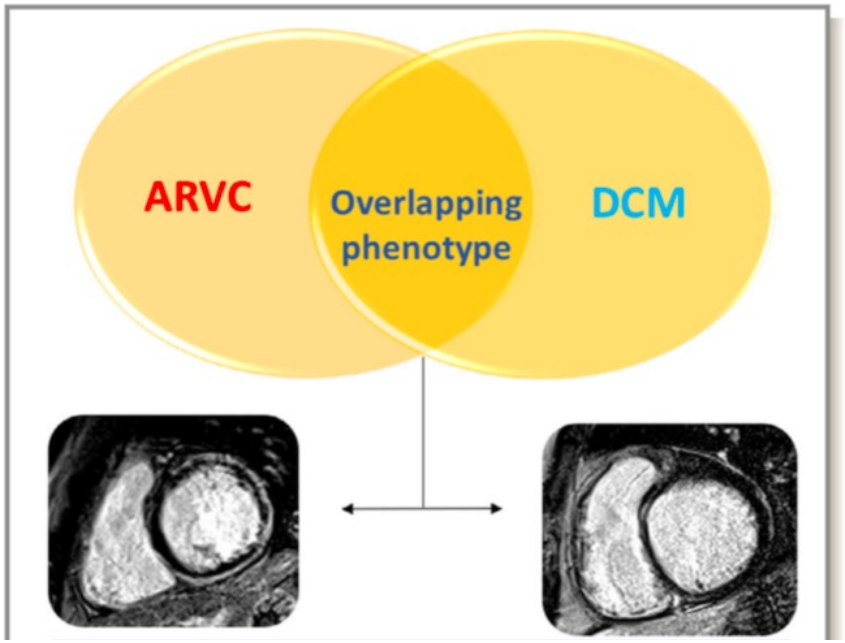
- Phenocopies ->mimicking LV-dominant

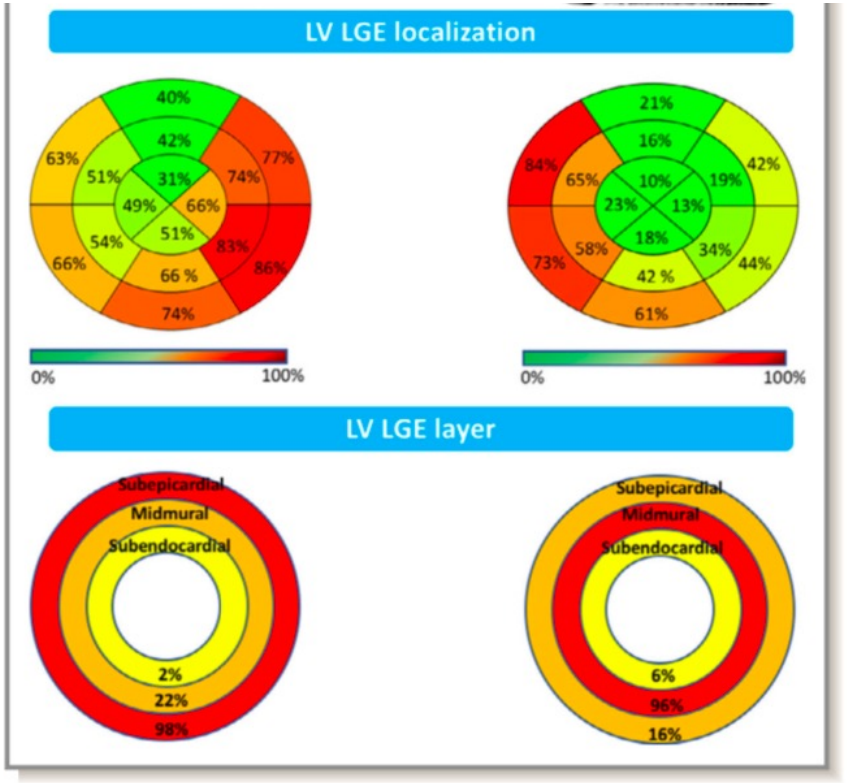


Dalla teoria alla pratica…

- La diagnosi differenziale della forma ARVC
- La diagnosi differenziale della forma ALVC
- La diagnosi differenziale nelle forme dilatative

Arrhythmogenic Right Ventricular Cardiomyopathy: Characterization of Left Ventricular Phenotype and Differential Diagnosis With Dilated Cardiomyopathy





Cipriani J Am Heart Assoc. 2020

Come distinguo una DCM da una cardiomiopatia aritmogena LV-dominant?

	CLINICA	ECG	ARITMIE	CMR
ALVC	Gene-related? -Sincope	- Bassi voltaggi nelle derivazioni perifieriche (<0.5 mV) - Onde T negative infero-laterali	TV monomorfa BBDx	- **LGE 100% dei casi** - Importante quantità di LGE (>20%) - Relazione diretta tra quantità di LGE e disfunzione sistolica - LVLGE con pattern subepicardico/midmural circolare >> infero-laterale laterale
DCM	Scompenso cardiaco; - Aritmie «gene-related»?	- IVS - BBSX		- **LGE 40-45%** - Poca quantità LGE (<10-15%) - Disfunzione sistolica indipendente dalla quantità di LGE - LV LGE >> SIV

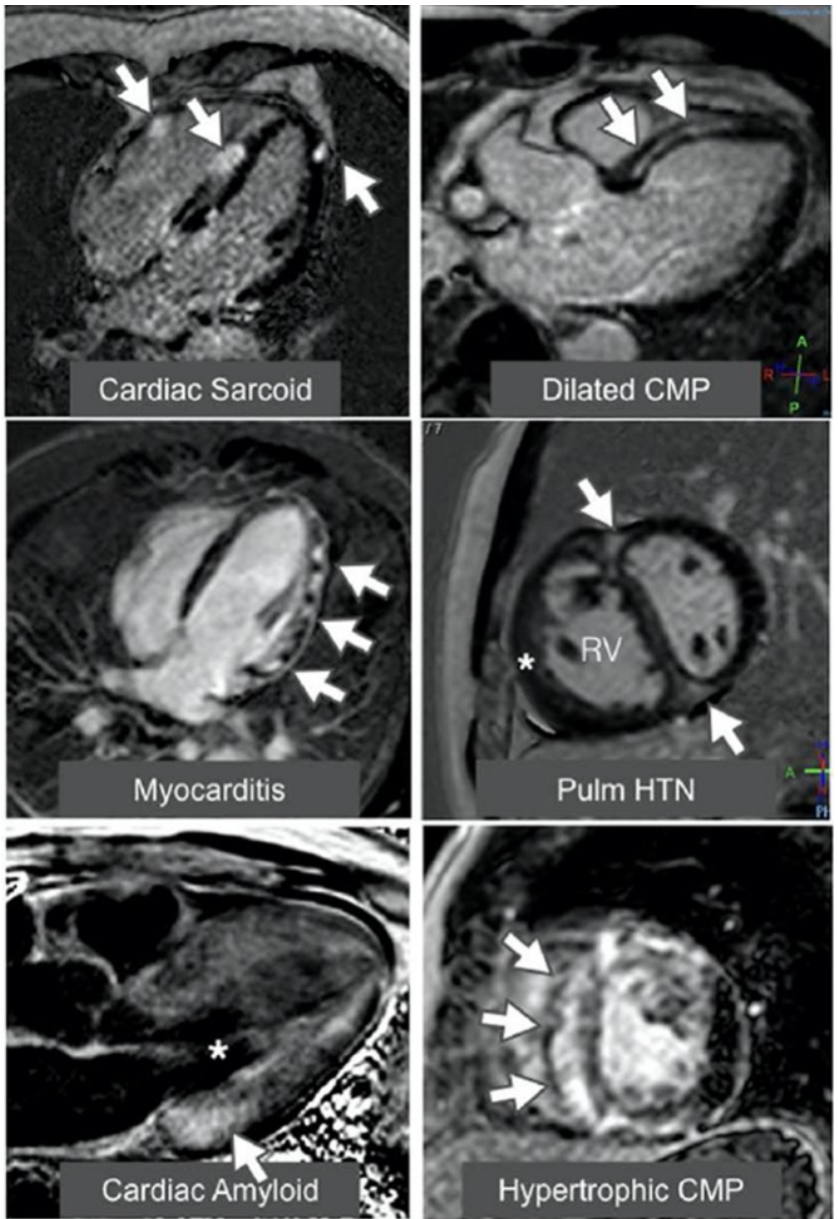
Protocollo di esecuzione

- CARDIOMIOPATIE PRIMARIE
  - ARVC

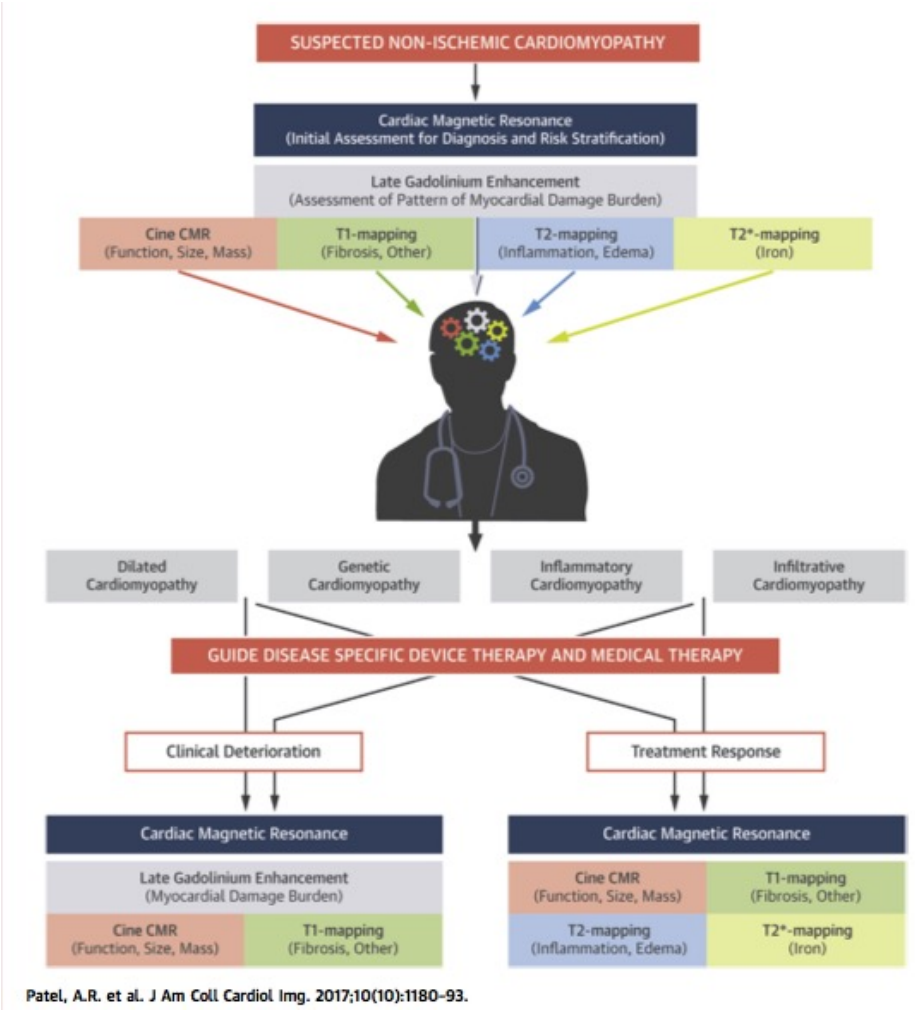
J Cardiovasc Med 2017

ARVC			
Cause of RV dilation or dysfunction	8	–	Cine-bSSFP in short-axis views from mitral valve to apex Q_p/Q_s evaluation with VEPC
Identification of biventricular tissue and wall motion abnormalities in patients with ventricular arrhythmias	7	–	Cine-bSSFP in short-axis and long-axis views FSE in short-axis and long-axis views LGE in short-axis and long-axis views
Follow-up CMR in patients with tissue abnormalities or wall motion abnormalities or RV dilatation, not satisfying TF criteria	7	–	As above
CMR in mutation carries or familiar of a ARVC proband	7	–	As above

In conclusione…



Role of Cardiac Magnetic Resonance in the Diagnosis and Prognosis of Non-Ischemic Cardiomyopathy



Patel, A.R. et al. J Am Coll Cardiol Img. 2017;10(10):1180–93.

Patel and Kramer 2017 JACC: Cardiovascular Imaging

GRAZIE DELL'ATTENZIONE

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