

Fenotipo ipertrofico update 2026

Ruolo della CMR nella HCM: oltre la quantificazione del LGE

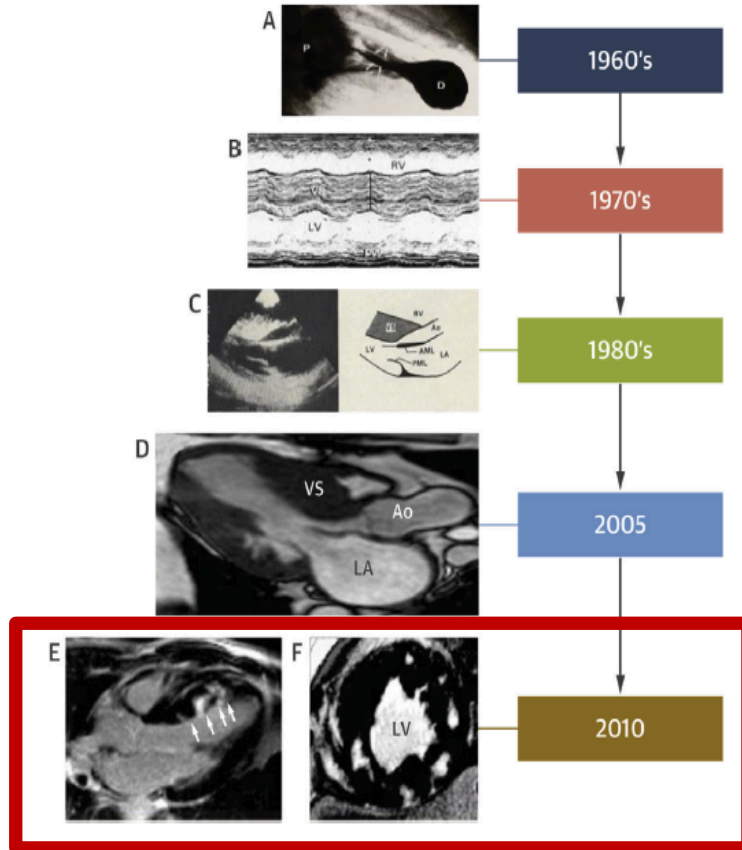
Giancarlo Todiere, MD PhD
FTGM Pisa



Fondazione
Monasterio
la ricerca che cura

Nessun conflitto di interesse

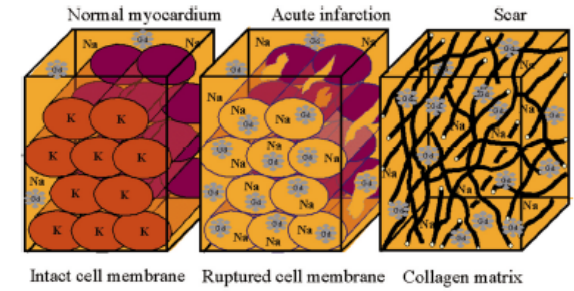
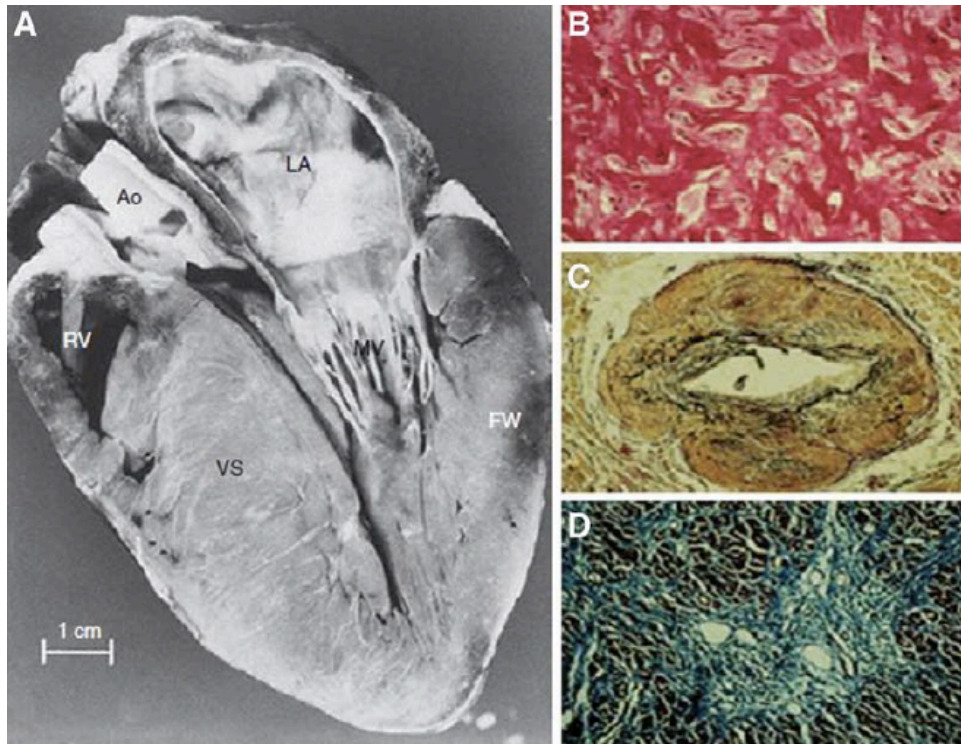




Miglioramento delle tecniche di Imaging

CMR

Maron BJ JACC 2016



Disarray, fibrosi, malattia piccoli vasi

Definizione ipertrofia e valutazione morfologia

Journal of the American College of Cardiology
© 2009 by the American College of Cardiology Foundation
Published by Elsevier Inc.

2009

Vol. 54, No. 3, 2009
ISSN 0735-1097/09/\$36.00
doi:10.1016/j.jacc.2009.05.006

FOCUS ISSUE: HYPERTROPHIC CARDIOMYOPATHY

Clinical Research

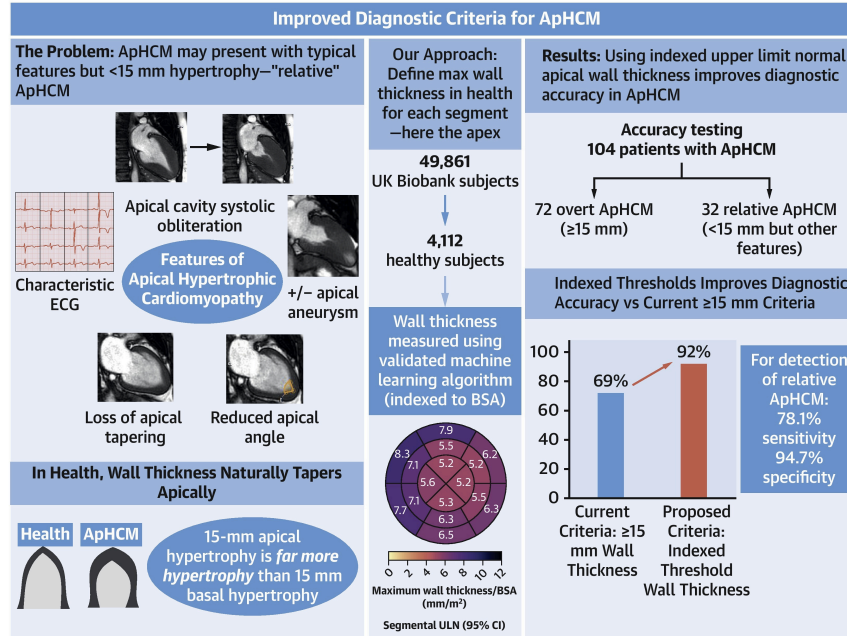
Hypertrophic Cardiomyopathy Phenotype Revisited After 50 Years With Cardiovascular Magnetic Resonance

Martin S. Maron, MD,* Barry J. Maron, MD,† Caitlin Harrigan, BA,* Jacki Buros, BA,‡
C. Michael Gibson, MD, MS,‡§ Iacopo Olivotto, MD,|| Leah Biller, BA,† John R. Lesser, MD,†
James E. Udelson, MD,* Warren J. Manning, MD,‡§ Evan Appelbaum, MD‡§

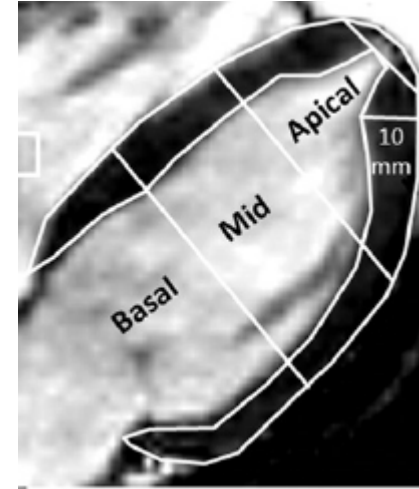
Boston, Massachusetts; Minneapolis, Minnesota; and Florence, Italy

12% non individuate

CENTRAL ILLUSTRATION: Using New Indexed Apical Wall Thickness Thresholds Improves the Diagnostic Accuracy of ApHCM >5.6 mm/m²



Hughes RK, et al. J Am Coll Cardiol Img. 2024;17(5):501-512.

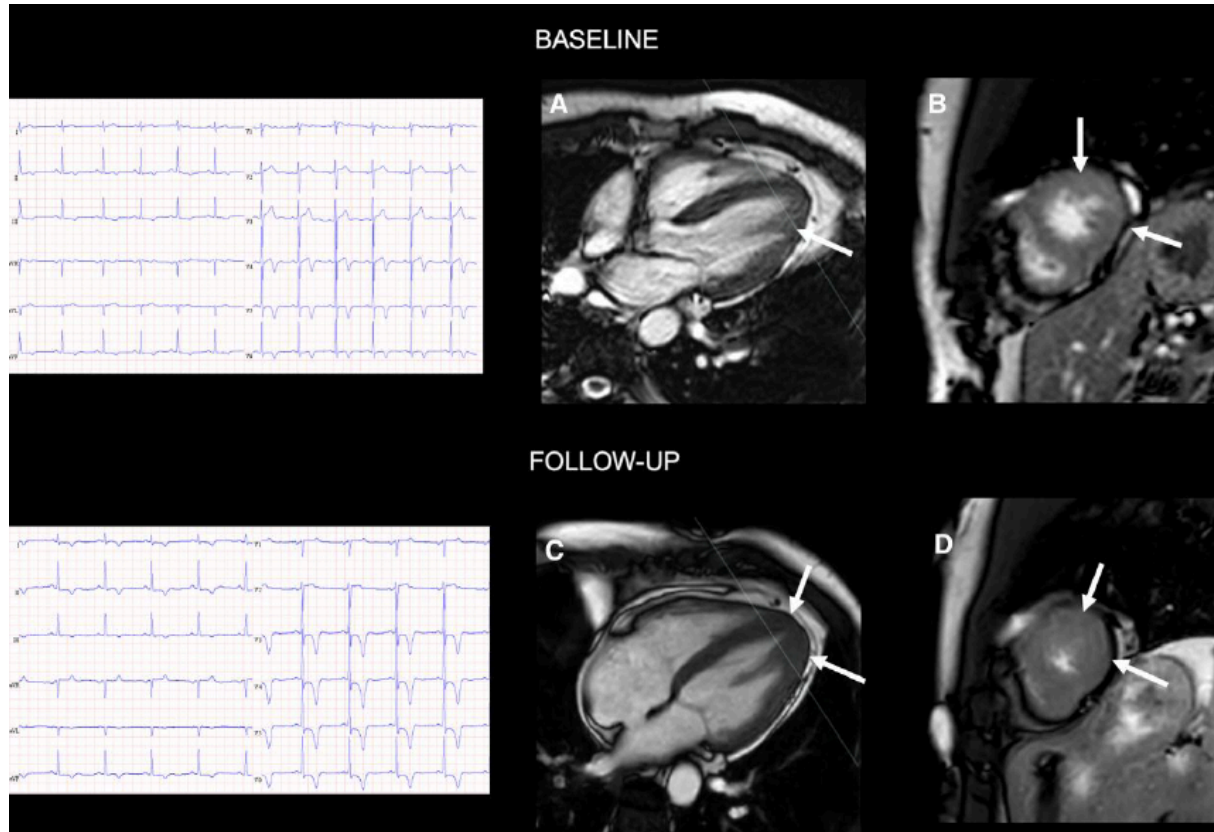


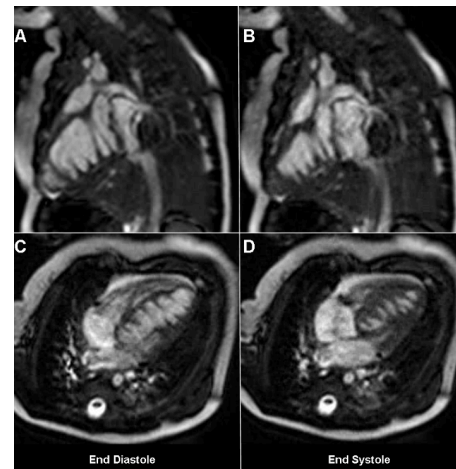
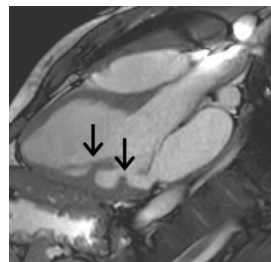
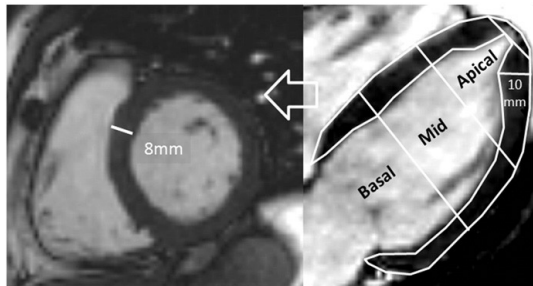
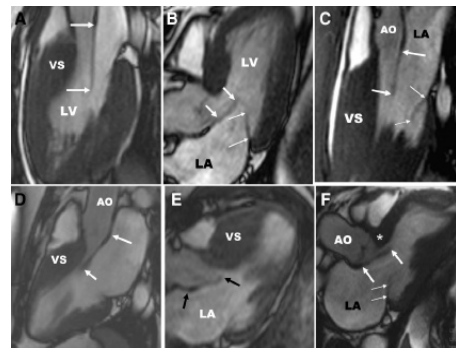
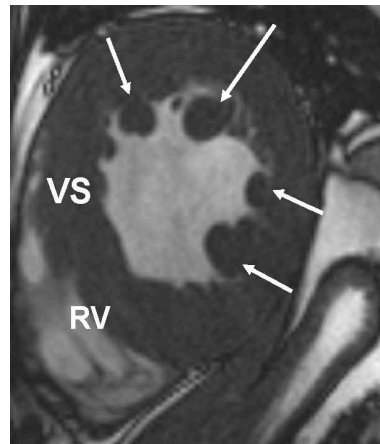
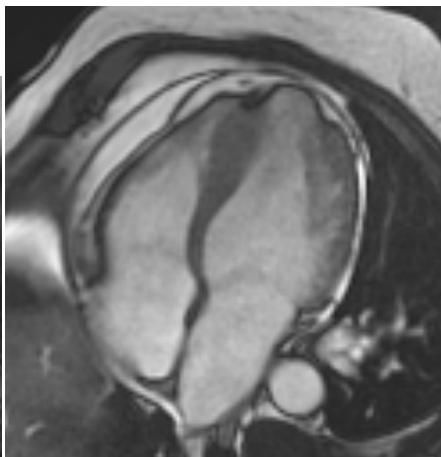
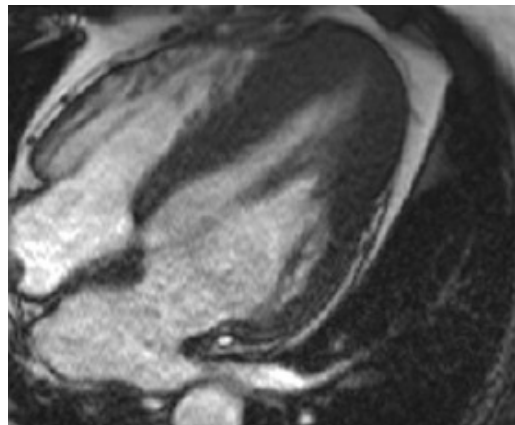
Ipertrofia relativa apicale
APICE/BASE >1

/LGE/ aneurismi apicali/ dilatazione atrio sn/
obliterazione sistolica apice

Flett AS, IJC 2015

Follow-up: HCM apicali



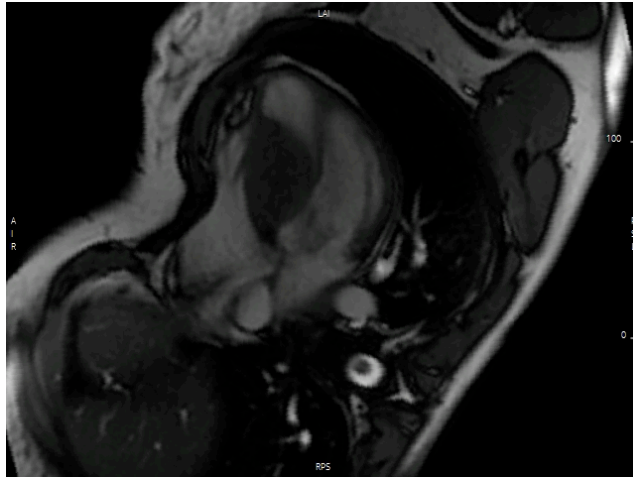


Ipercontrattilità & dispendio energetico

Circulation. 2021 November 23; 144(21): 1714–1731. doi:10.1161/CIRCULATIONAHA.121.053575.

Altered Cardiac Energetics and Mitochondrial Dysfunction in Hypertrophic Cardiomyopathy

Sara Ranjbarvaziri, PhD^{1,2}, Kristina B. Kooiker, PhD³, Mathew Ellenberger, BS⁴, Giovanni Fajardo, MD^{1,2}, Mingming Zhao, MD^{1,2}, Alison Schroer Vander Roest, PhD^{1,2}, Rahel A. Woldeyes, PhD⁵, Tiffany T. Koyano, BS⁶, Robyn Fong, BS⁶, Ning Ma, PhD^{2,7}, Lei Tian, PhD^{2,7}, Gavin M. Traber, BS⁴, Frandies Chan, MD⁸, John Perrino, BS⁹, Sushma Reddy, MD^{1,2}, Wah Chiu, PhD^{5,10}, Joseph C. Wu, MD, PhD^{2,7}, Joseph Y. Woo, MD⁶, Kathleen M. Ruppel, MD^{1,11}, James A. Spudich, PhD¹¹, Michael P. Snyder, PhD⁴, Kévin Contrepois, PhD⁴, Daniel Bernstein, MD^{1,2}



EF 70%

Cardiac MRI (al massimo potenziale)

Diagnosi eziologica???



Cardiomyopathy
phenotype

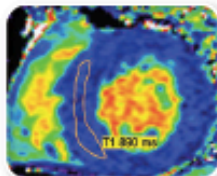
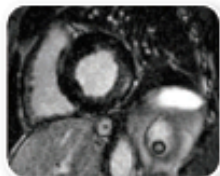
Finding

Cardiac CMR examples

Specific diseases to be considered

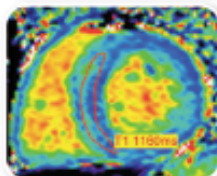
HCM

Posterolateral LGE and concentric LVH
Low native T1



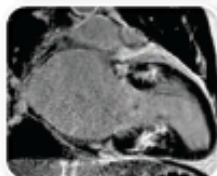
Anderson–Fabry disease

Diffuse subendocardial LGE,
high native T1



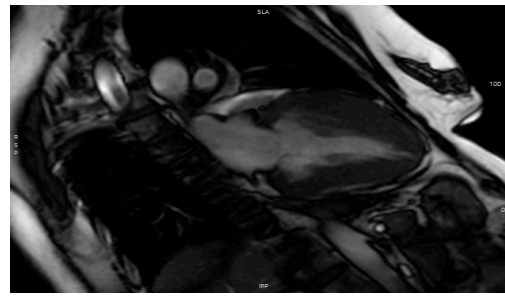
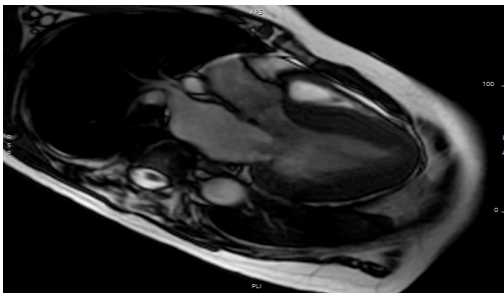
Amyloidosis

Patchy mid-wall in hypertrophied areas



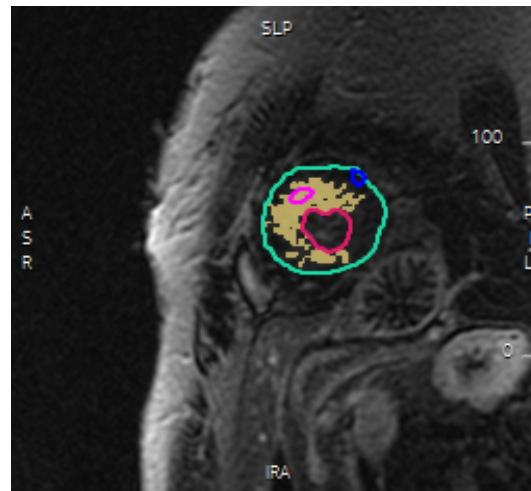
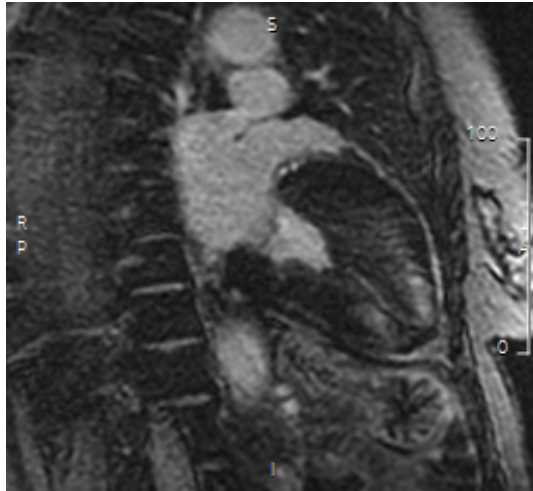
Sarcomeric HCM

ESC guidelines 2023

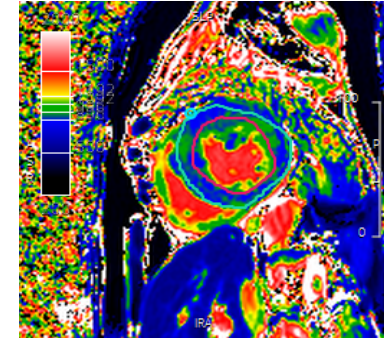


I CASO

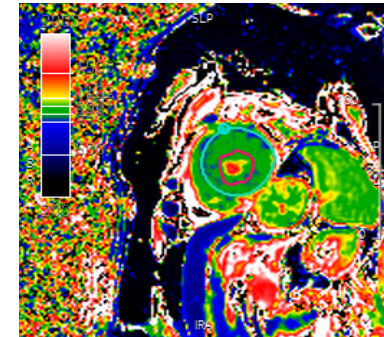
Ipertrofia concentrica



LGE setto-apice circa 11% (6 SD metodo)

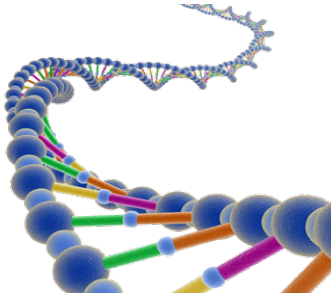
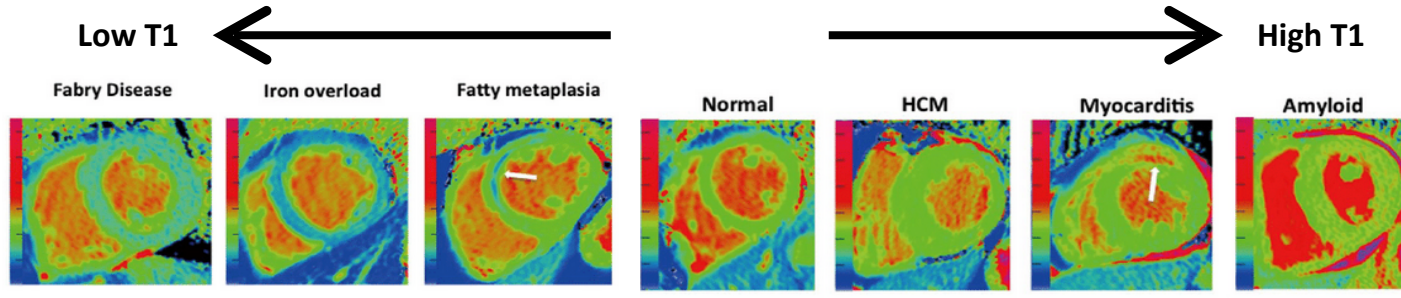


T1 nativo basale ridotto 935 ms



T1 apice pseudonormale 990 ms

T1 nativo



Da mutazione VUS MYBPC3

Mutazione

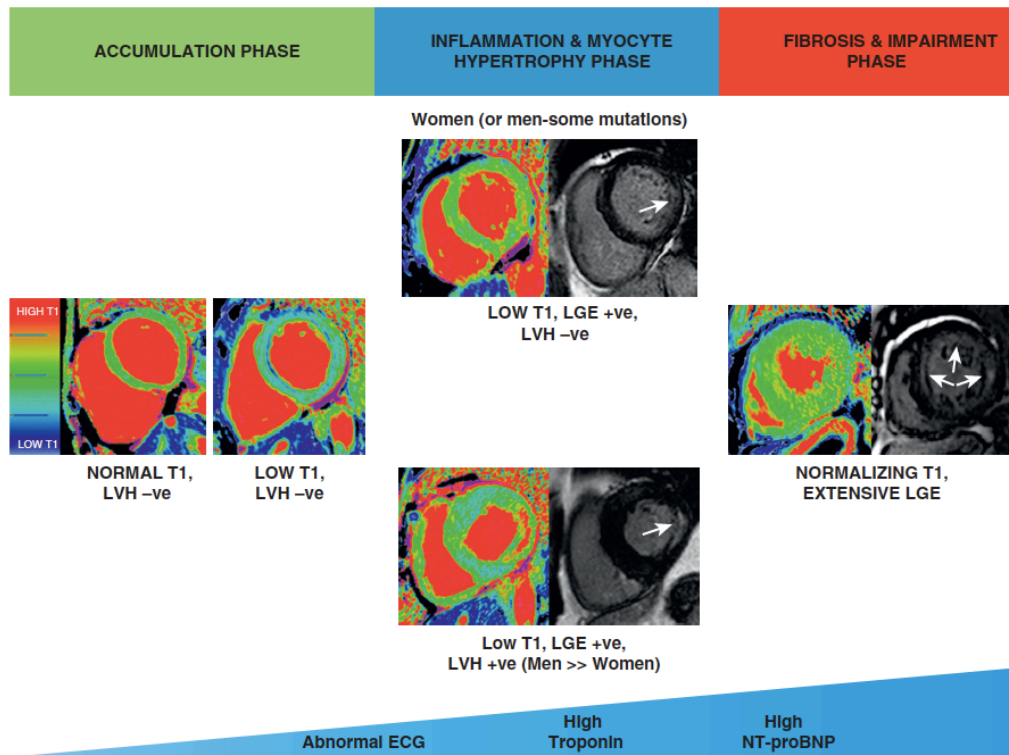
c.548-3_553 del per GLA

(splice junction loss)

Malattia di FABRY

2 figli di cui una portatrice della stessa mutazione

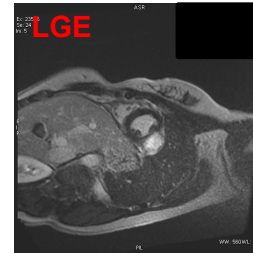
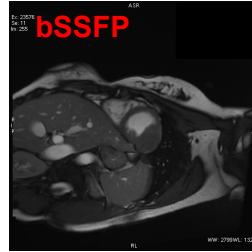




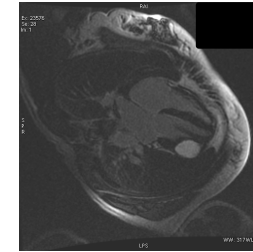
II CASO

Ipertrofia parete laterale

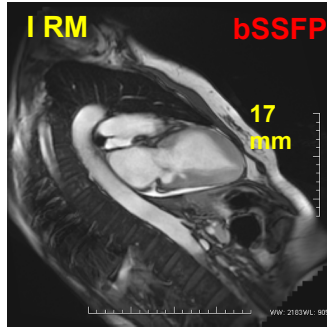
Sequenze T1W e T2W



- Adolescenti
- Intramurale setto
- Rischio elevato Sudden Death

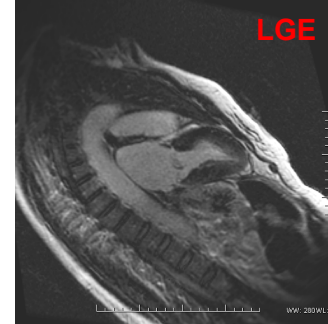
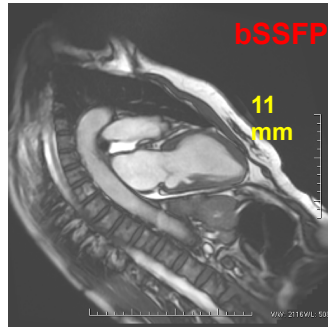


Fibroma



**Sospetto
Takotsubo**

II RM



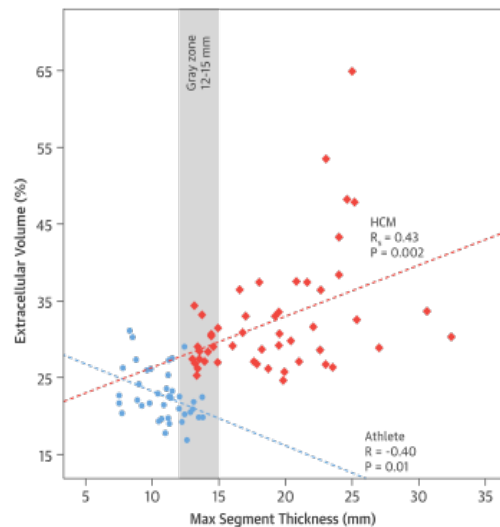
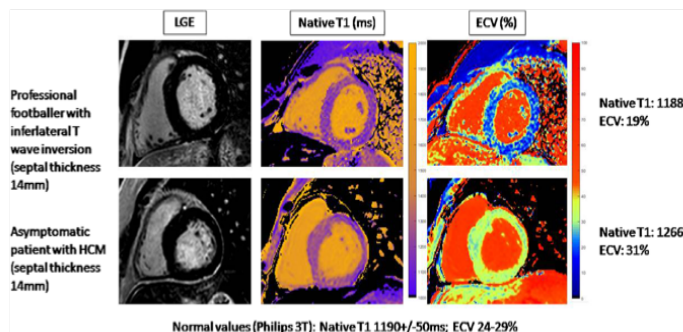
III CASO
Ipertrofia apicale

Sequenze T2W

Volumi, massa, ECV

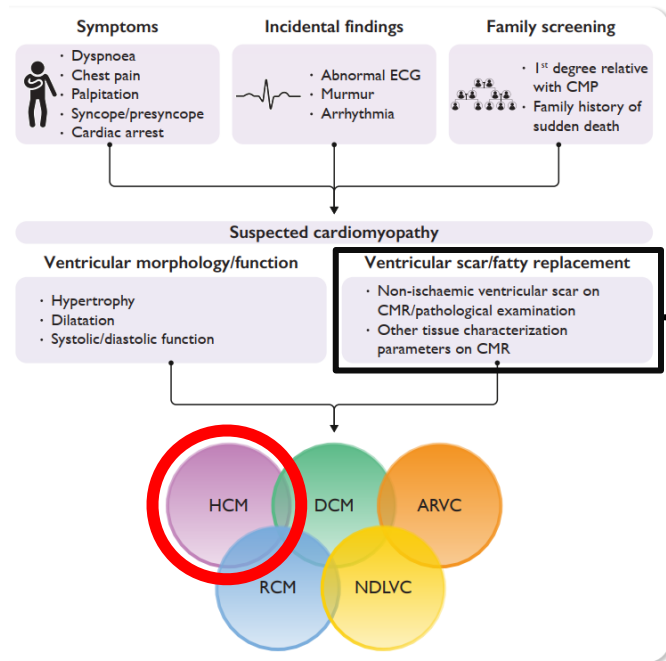
Assessing Myocardial
Extracellular Volume
by T1 Mapping to
Distinguish Hypertrophic
Cardiomyopathy From
Athlete's Heart

Cuore d'Atleta



JACC 2016

PROGNOSIS/FOLLOW UP



Ventricular scar/fatty replacement

- Non-ischaemic ventricular scar on CMR/pathological examination
- Other tissue characterization parameters on CMR

Solo LGE quantitativo???

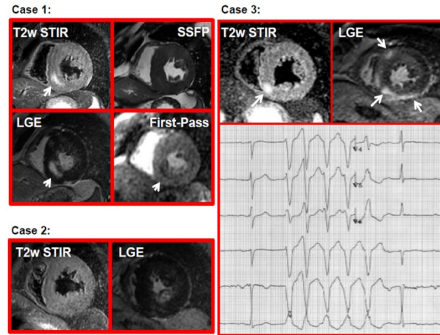
T2 imaging: qualitativo

OPEN ACCESS Freely available online

PLOS ONE

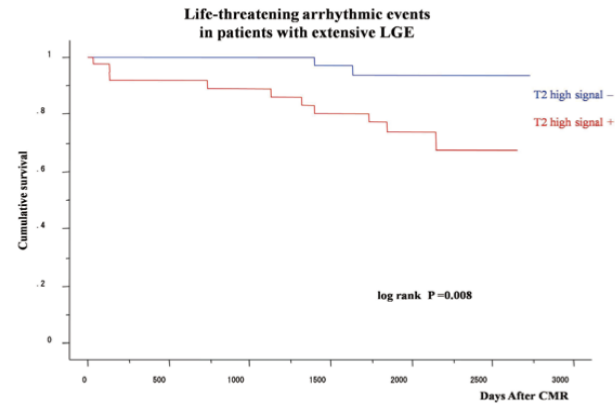
Abnormal T2-STIR Magnetic Resonance in Hypertrophic Cardiomyopathy: A Marker of Advanced Disease and Electrical Myocardial Instability

Giancarlo Todiere^{1,3}, Lorena Piscicelli², Andrea Barison¹, Annamaria Del Franco¹, Elisabetta Zachara², Paolo Piaggi³, Federica Re², Alessandro Pingitore⁴, Michele Emdin¹, Massimo Lombardi⁵, Giovanni Donato Aquaro^{1,3}

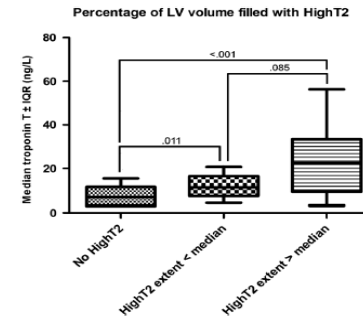


Independent variables	Logistic regression Analysis					
	NSVT	No NSVT	p value	Coefficient	Odds Ratio	95%-C.I.
Arrhythmic Risk factors	0.6±0.7	0.14±0.3	0.001			
LV mass index	131±38	103±39	0.007			
HtT2	21(95)	1(5)	<0.001	5.1	165	11–2455
Maximal Wall Thickness	25±6	19±6	<0.001	0.12	1.1	1.0–1.3
Extent of LGE	18.3±11.9	6.8±7.9	<0.001			

PlosONE 2014



Hen y et al Circ J 2018



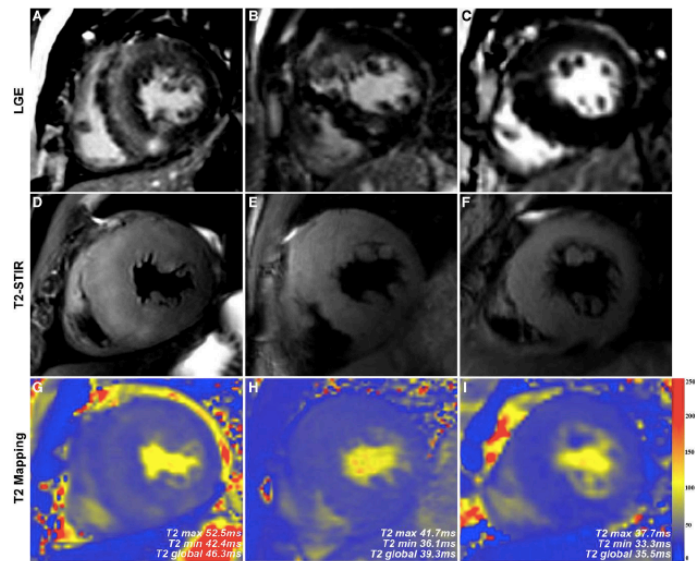
Danno miocardico

Gommans DHF Heart 2016

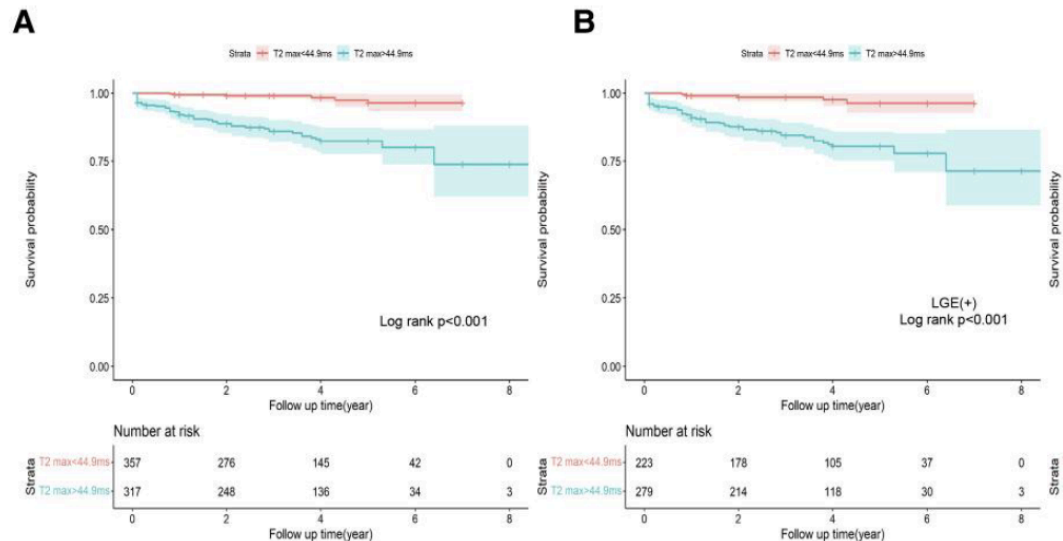
T2 imaging: quantitative

Incremental significance of myocardial oedema for prognosis in hypertrophic cardiomyopathy

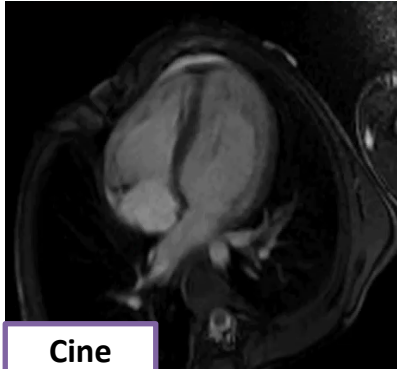
Ziqian Xu^{1,2}, Jie Wang², Wei Cheng¹, Ke Wan³, Weihao Li², Lutong Pu², Yuanwei Xu², Jiayu Sun¹, Yuchi Han ⁴, and Yucheng Chen ^{2*}



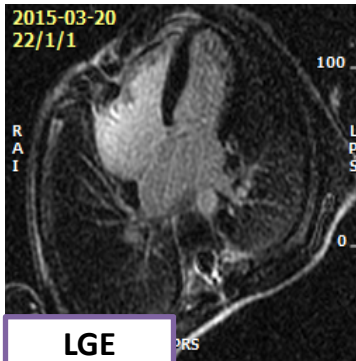
T2 MAPPING



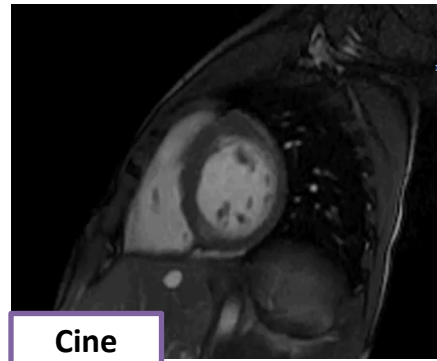
I RM 2015



Cine
SSFP



LGE

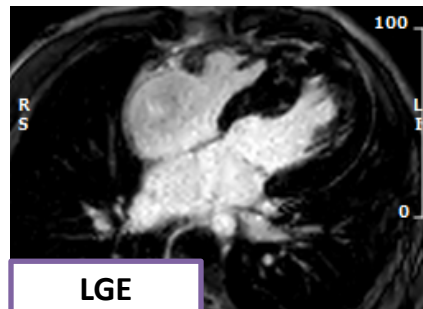


Cine
SSFP

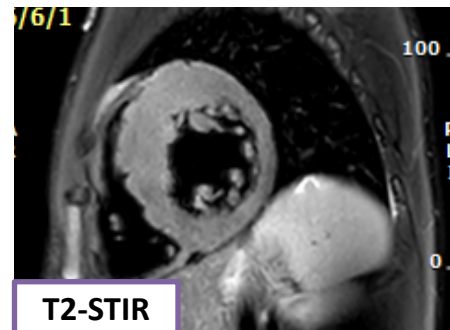
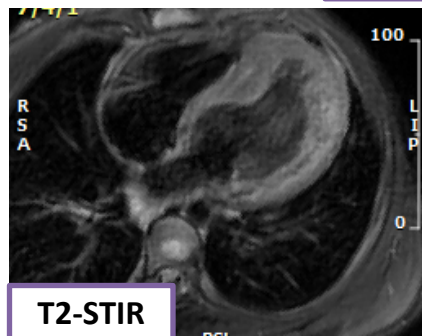
Ipertrabecolatura parete

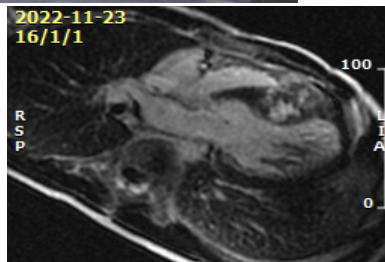
start
ATENOLOLO

II RM febbraio 2022

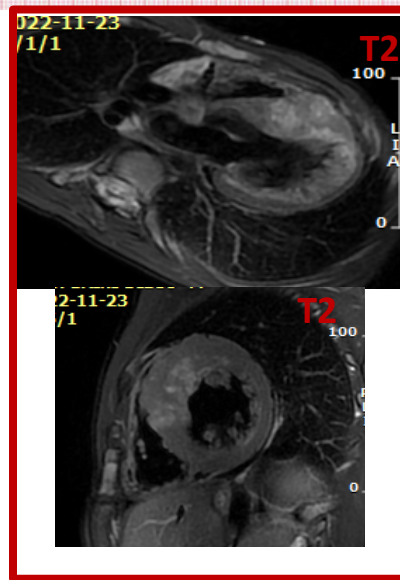


LGE (6SD) < 15%





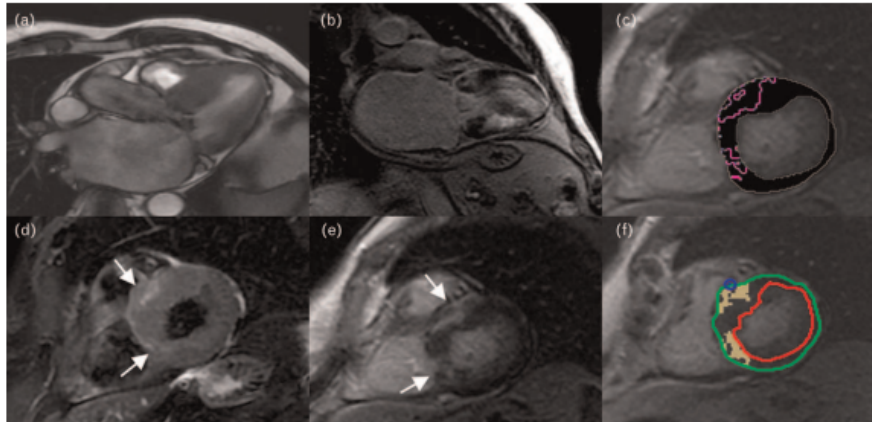
HOT-PHASE



Novembre 2022

Acute clinical presentation of nonischemic cardiomyopathies: early detection by cardiovascular magnetic resonance

Giancarlo Todiere^{a,*}, Andrea Barison^{a,*}, Anna Baritussio^b, Alberto Cipriani^b,
Andrea Igoren Guaricci^c, Silvia Pica^{d,†}, Ciro Indolfi^e, Gianluca Pontone^f and
Santo Dellegrottaglie^g, on behalf of the Working Group on Cardiac Magnetic
Resonance of the Italian Society of Cardiology



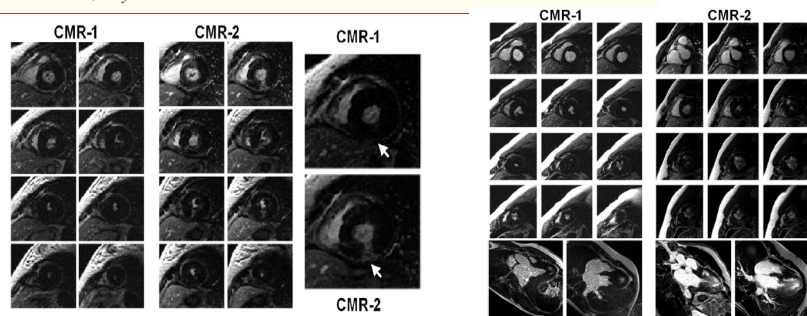
HOT PHASES!!!

Progression of Myocardial Fibrosis Assessed With Cardiac Magnetic Resonance in Hypertrophic Cardiomyopathy

JACC 2012

Giancarlo Todiere, MD,* Giovanni Donato Aquaro, MD,* Paolo Piaggi, MS, PhD,†
 Francesco Formisano, MD,‡ Andrea Barison, MD,§ Pier Giorgio Masci, MD,* Elisabetta Strata, MD,*
 Lorenzo Bacigalupo, MD,‡ Mario Marzilli, MD,|| Alessandro Pingitore, MD, PhD,¶
 Massimo Lombardi, MD*

Pisa and Genova, Italy



JACC: CARDIOVASCULAR IMAGING
 © 2016 BY THE AMERICAN COLLEGE OF CARDIOLOGY FOUNDATION
 PUBLISHED BY ELSEVIER

VOL. 19, NO. 6, 2016

ORIGINAL RESEARCH

2026

Serial Myocardial Fibrosis Assessments Predict Outcomes in Patients With Hypertrophic Cardiomyopathy



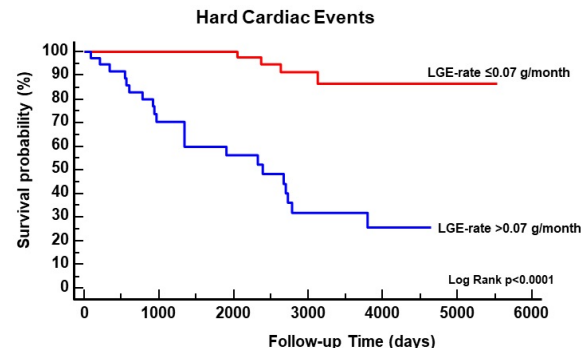
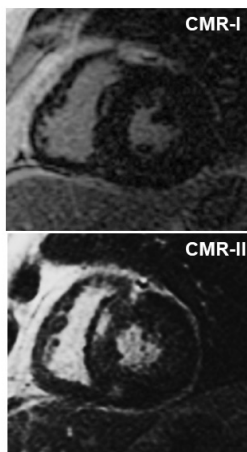
Yun Tang, MD,^{3,*} Xingrui Chen, MD,^{3,*} Kankan Zhao, MD,³ Xuan Ma, MD,³ Zhixiang Dong, MD,^{3,c} Jiabin Wang, MD,^c
 Bo Li, MD,³ Wenqing Xu, MD,³ Wenhao Dong, MD,³ Zhuxin Wei, MD,³ Xi Jia, MD,³ Pengyu Zhou, MD,³
 Yujie Liu, MD,³ Yanyan Song, MD,³ Chen Cui, MD,³ Kai Yang, MD,³ Xiuyu Chen, MD,³ Lei Xu, MD,³ Xiaohu Li, MD,³
 Yucheng Chen, MD,³ Lianming Wu, MD,³ Hui Liu, MD,³ Liming Xia, MD,³ Shujuan Yang, MD,³
 Shihua Zhao, MD, PhD³†

LGE progressivo (quantitativo ma valutato nel tempo)



105 pts

52 mesi di
 follow-up



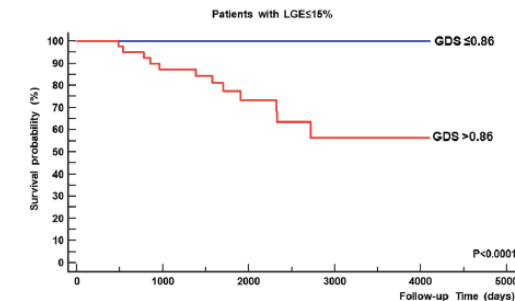
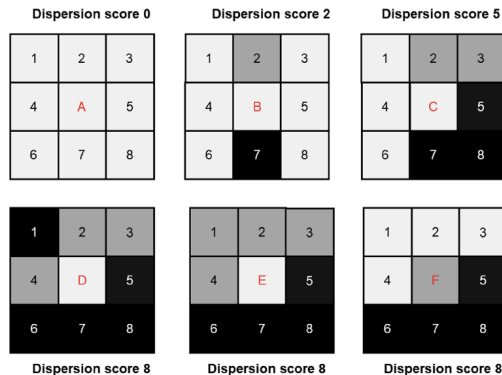
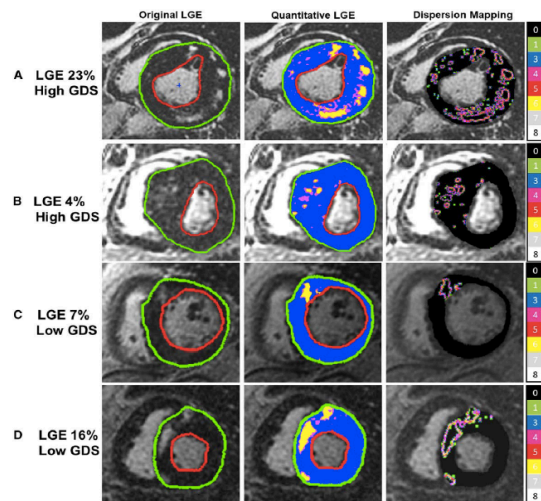
	Number at risk					
LGE-rate ≤0.0768	62	42	22	6	1	0
LGE-rate >0.0737	22	16	7	2	0	0

Todiere G , Aquaro GD, American Journal of Cardiology 2023

ORIGINAL ARTICLE

Late Gadolinium Enhancement–Dispersion Mapping

A New Magnetic Resonance Imaging Technique to Assess Prognosis in Patients With Hypertrophic Cardiomyopathy and Low-Intermediate 5-Year Risk of Sudden Death



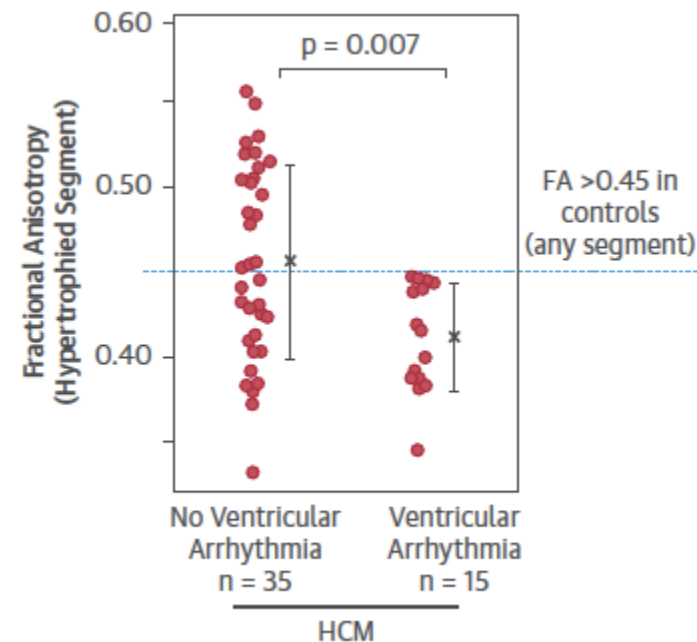
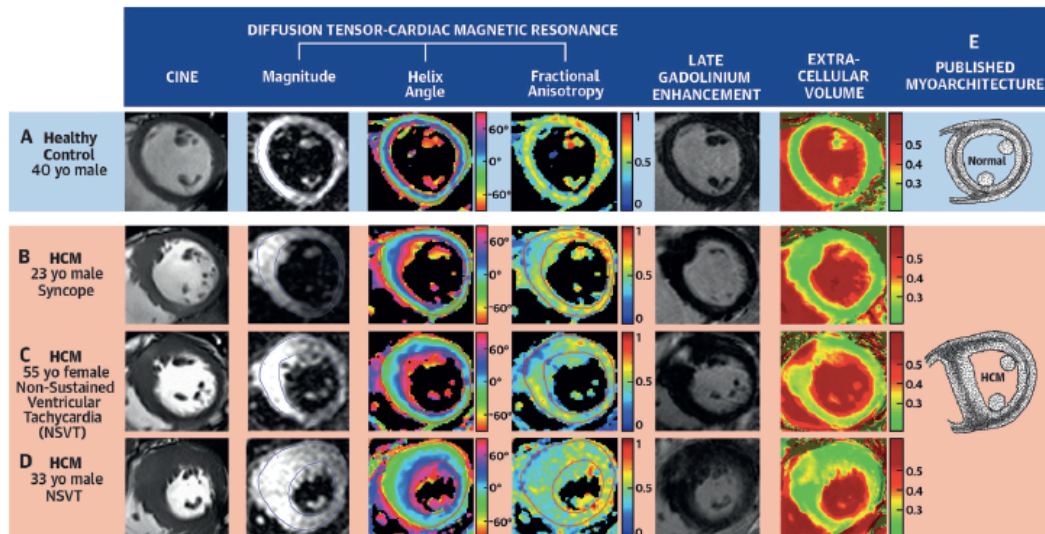
Number at risk:

GDS ≤ 0.86	112	85	47	19	4	0
GDS > 0.86	42	33	17	8	3	0

Eterogeneità Fibrosi

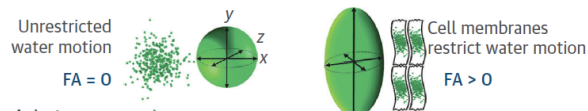
Aquaro GD et al. 2021

Non solo fibrosi...

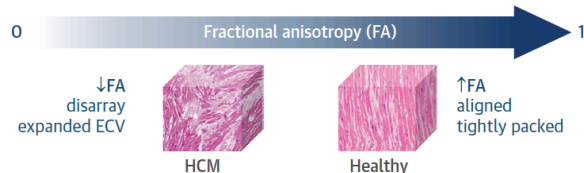


DISARRAY

Diffusion Tensor Cardiac Magnetic Resonance maps 3D motion of water

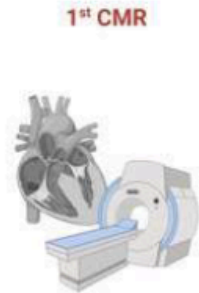


Anisotropy:
from Greek *anisos* "unequal" + *tropos* "turn" = "not the same in all directions"



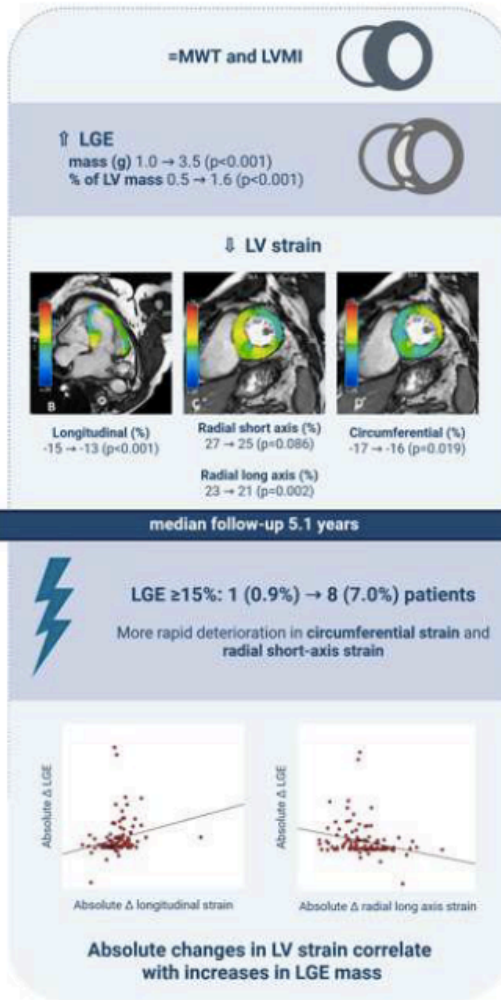
Ariga R et al. JACC 2019

Strain

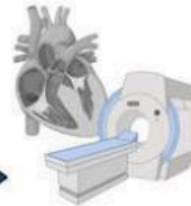


1st CMR

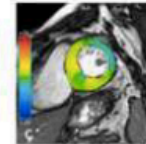
114 HCM patients
median age 51 y.o.
28% women
14% oHCM



2nd CMR



In patients with baseline LGE <15%
yearly absolute changes in
longitudinal ($p = 0.035$),
radial long-axis ($p = 0.014$)
and radial short-axis strain ($p = 0.006$)
are predictors of adverse CV events



Radial short-axis strain predictive even on top of HCM score and LGE extent at baseline ($p = 0.011$)

40 (35%) patients



Composite endpoint



SCD



Life-threatening VAs



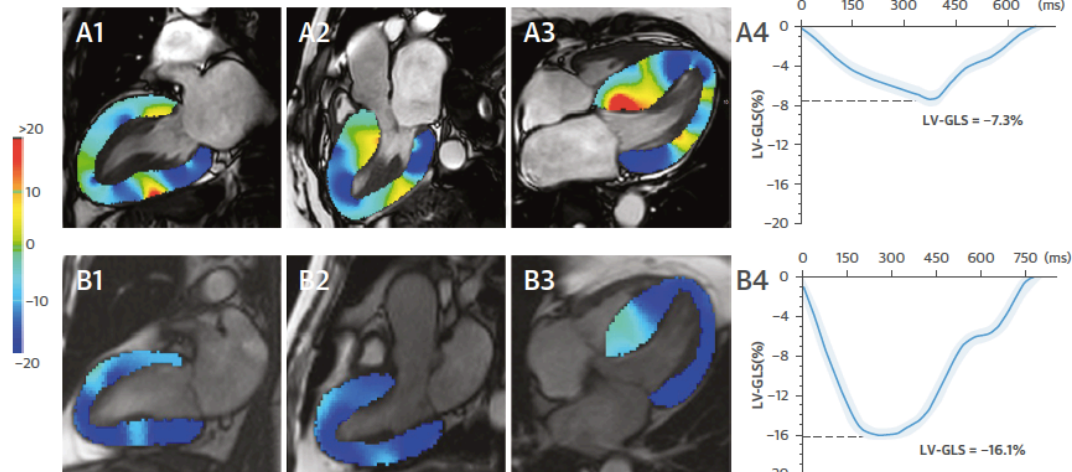
Stroke



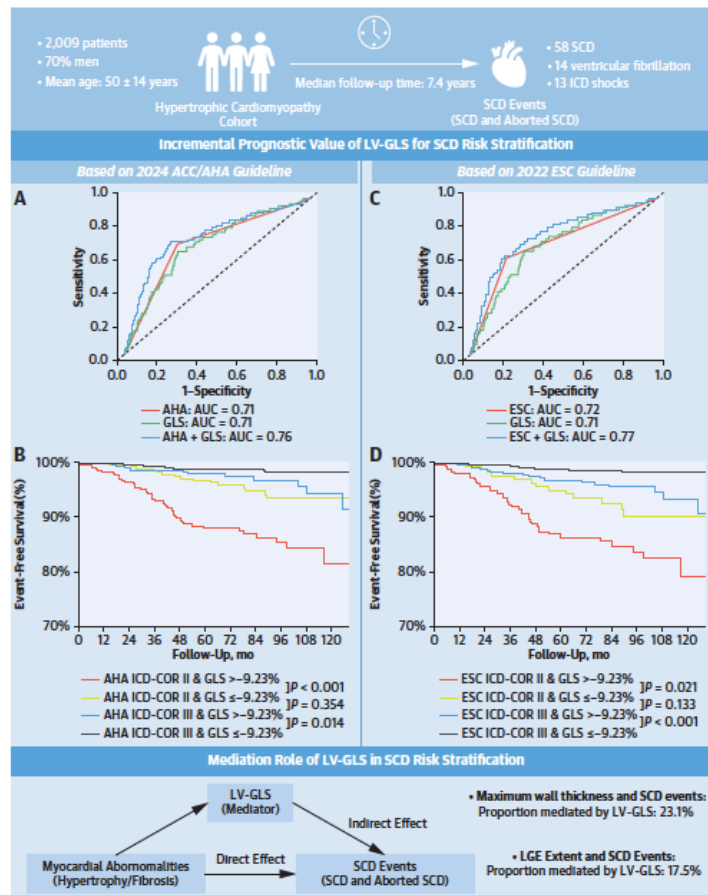
New-onset AFib

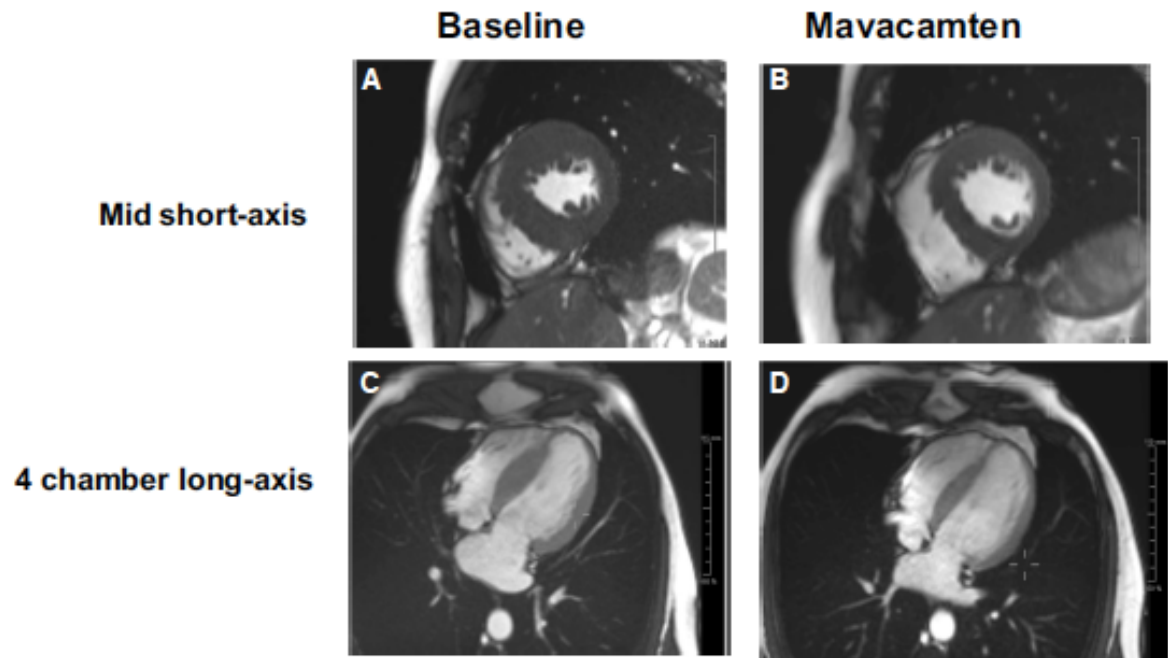


HF hospitalization



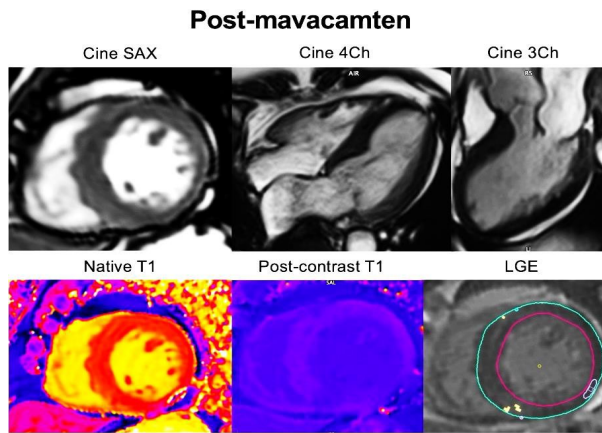
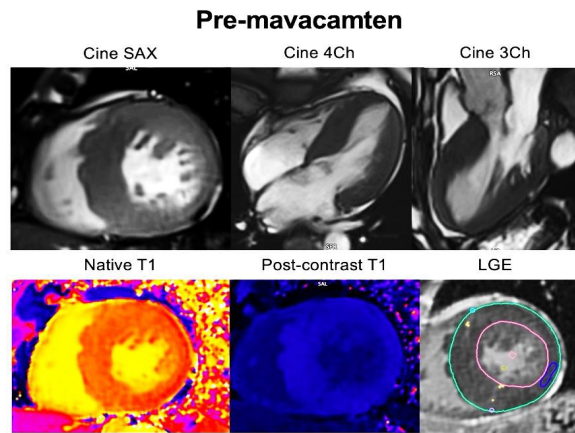
- Disarray
- Ischemia miocardica/
anomalie
energetiche
- edema





Saberi S et al. Circulation 2021

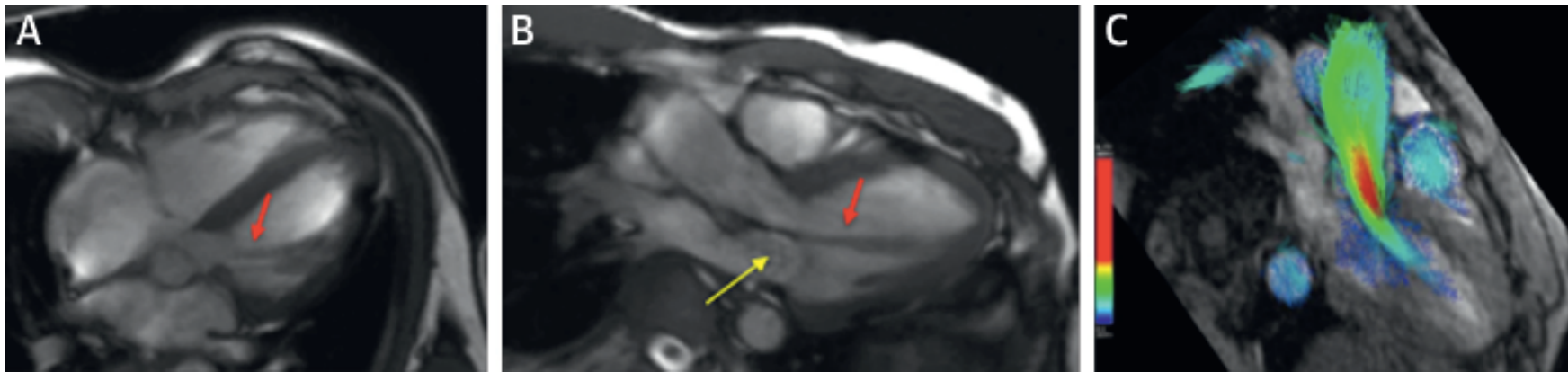
MEMENTO! Long term study



1-ECV

Characteristic	Pre-mavacamten	Post- mavacamten	Post - Pre Difference
BSA-indexed LV mass, g/m ²	113.6	71.5	-42.1
ECV fraction	0.30	0.33	0.03
BSA-indexed Cellular volume, mL/m ²	79.8	47.7	-32.0
BSA-indexed Matrix volume, mL/m ²	33.8	23.8	-10.0
LGE mass by 6SD, g	0.29	0.39	0.10

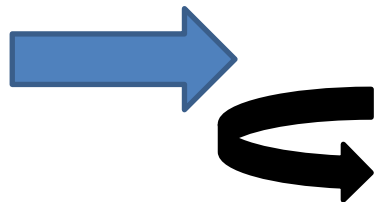
4D Flow imaging



Sherrid MV

1980

ECHO
Funzione



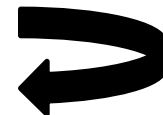
2000

RM
ECG
Tessuto



2020

RM
ECHO
Funzione



Rapporto
**Tessuto/
Funzione**

TAKE HOME MESSAGES

RM solo LGE quantitativo???

- ✓ **RM è fondamentale nella diagnosi delle cardiomiopatie a fenotipo ipertrofico (T1w-T2w)**
- ✓ **Ruolo nella prognosi delle cardiomiopatie (T2w, eterogeneità e progressione LGE)**
- ✓ **Esame senza radiazioni ionizzanti, ripetibile nel tempo ogni 3-5 anni, utile nel follow-up**
- ✓ **Con l'avvento degli «inibitori della miosina» potenzialità nella valutazione dell'efficacia delle terapie**



Grazie per l'attenzione
gtodiare@ftgm.it