

PLACE



PLATFORM OF LABORATORIES FOR ADVANCES IN CARDIAC EXPERIENCE

ROMA

Centro Congressi
di Confindustria

**Auditorium
della Tecnica**

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1 Ottobre

2022



CARDIOMIOPATIA IPERTROFICA: UPDATE 2022

Giancarlo Todiere, MD, PhD

Fondazione Toscana Gabriele Monasterio-Pisa

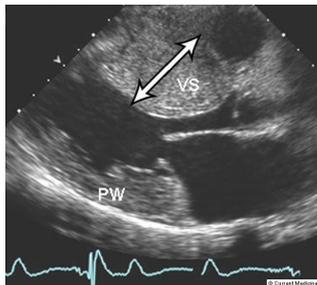


RM CARDIOMIOPATIA IPERTROFICA



Distribuzione ed estensione dell'ipertrofia ventricolare?

Diagnosi differenziale tra i diversi fenotipi di ipertrofia?



Mark
sfa



ione clinica

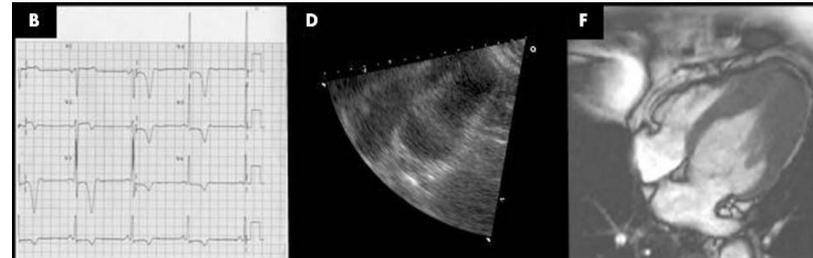


Journal of the American College of Cardiology Vol. 54, No. 3, 2009
 © 2009 by the American College of Cardiology Foundation
 Published by Elsevier Inc. ISSN 0735-1097/09/\$36.00
 doi:10.1016/j.jacc.2009.05.006

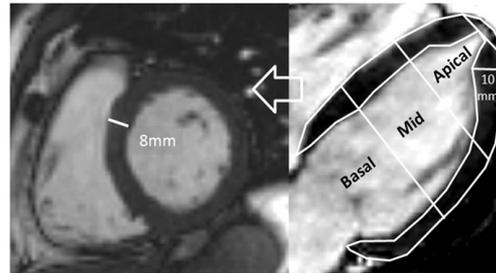
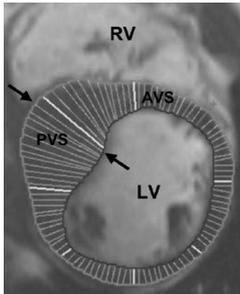
FOCUS ISSUE: HYPERTROPHIC CARDIOMYOPATHY **Clinical Research**

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 Olivetto, 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

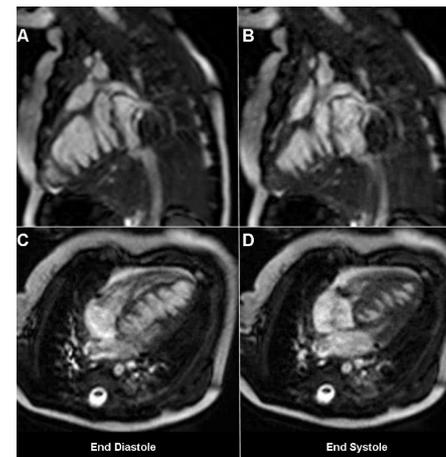
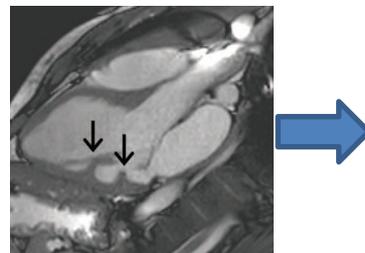
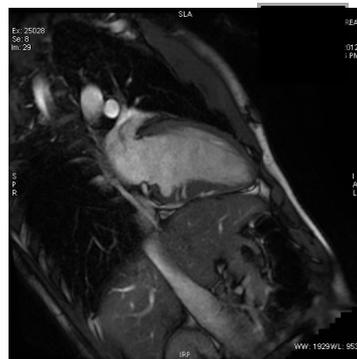
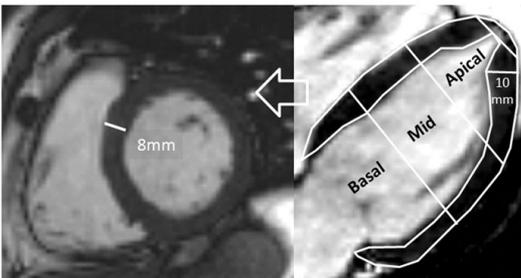
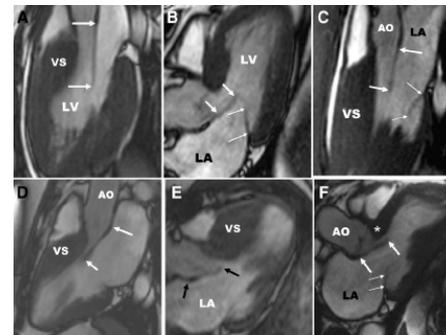
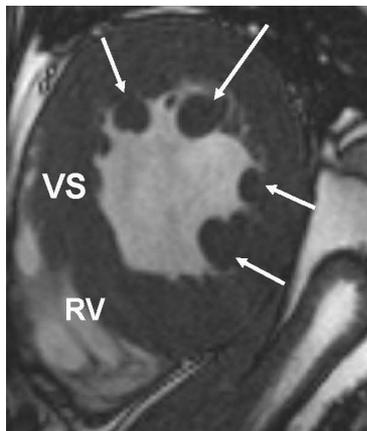
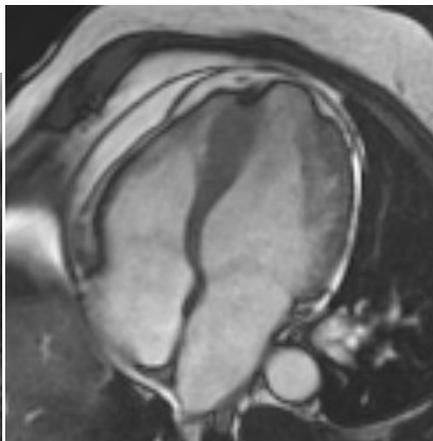
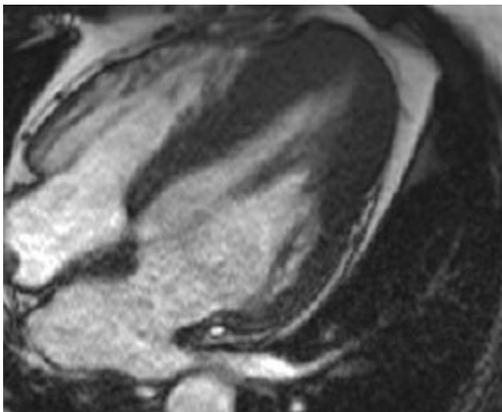


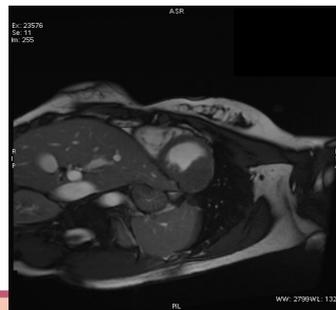
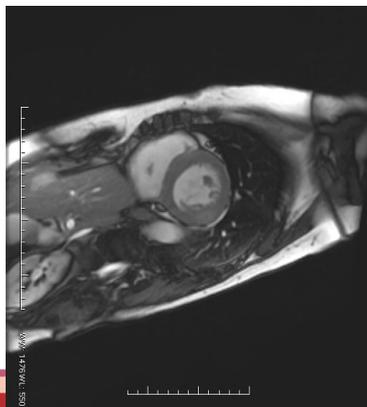
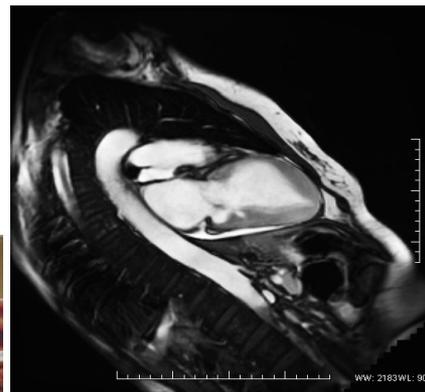
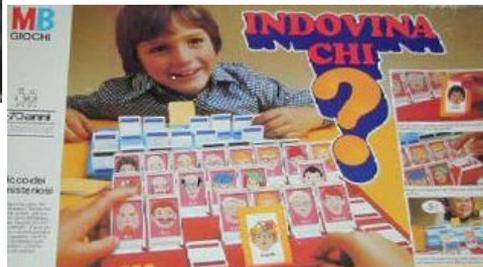
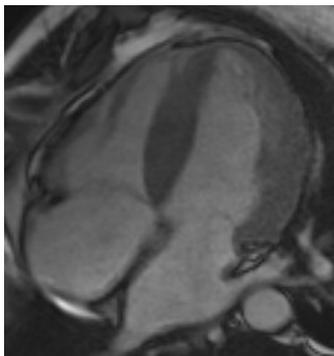
Ipertrofia relativa apicale

APICE/BASE >1

Flett AS et al. IJC 2015

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

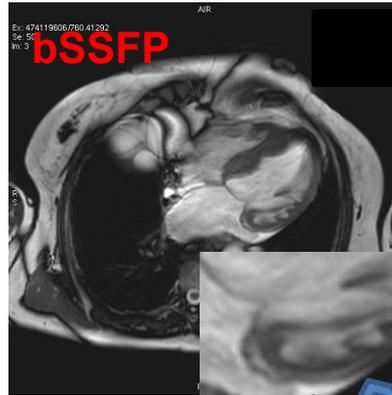






I CASO

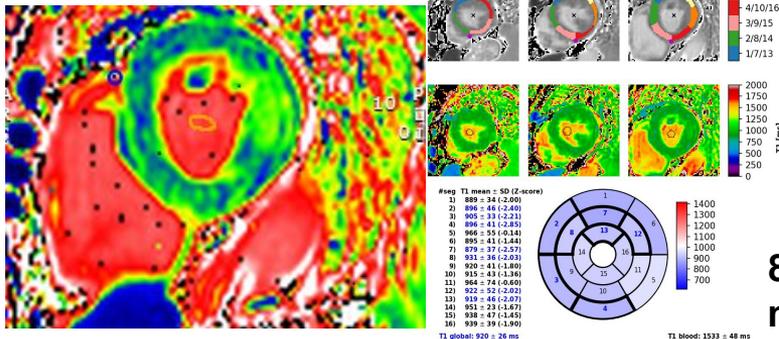
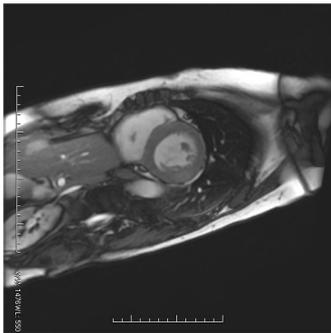
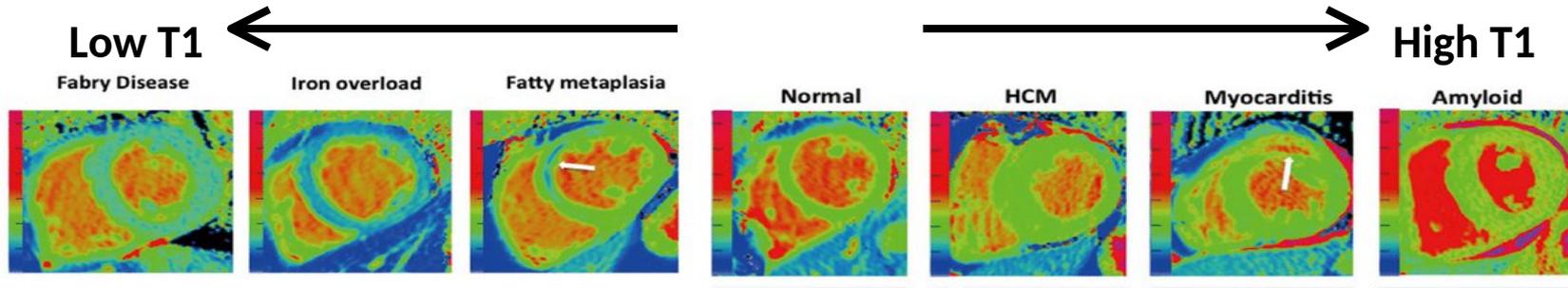
Ipertrofia asimmetrica VS 20 mm



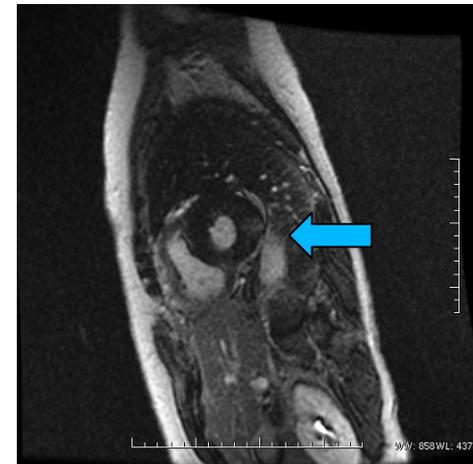
**CARDIOMIOPATIA IPERTROFICA
mutazione MYBPC3**



Malattia di Fabry



879 msec



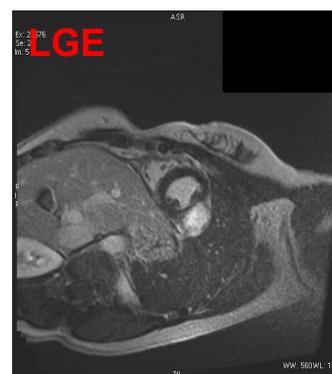
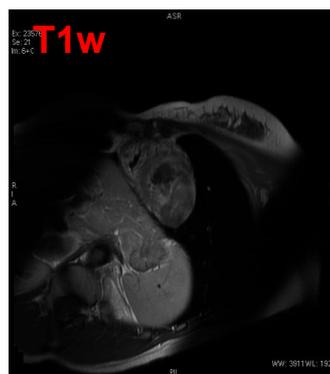
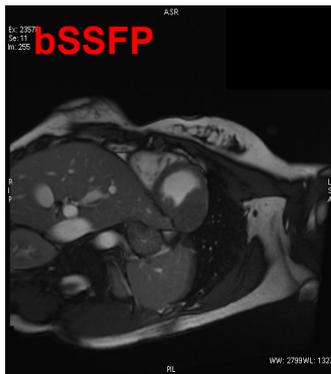
II CASO

SIV 14 mm, PL 15 mm, MASSA 124 gr

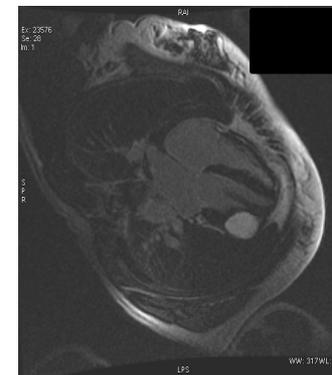
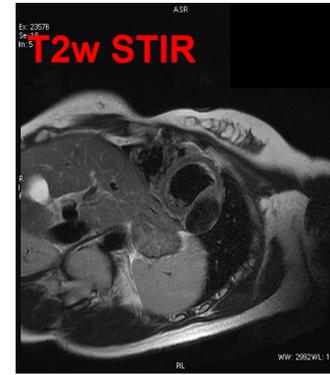


III CASO

Fibroma



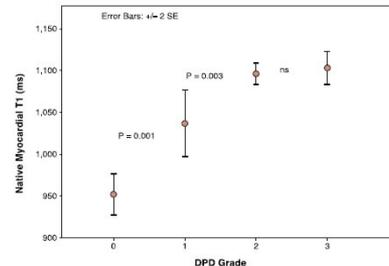
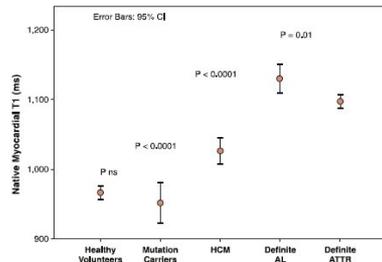
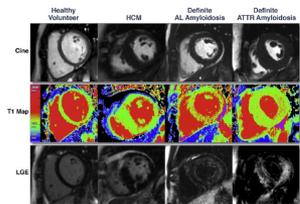
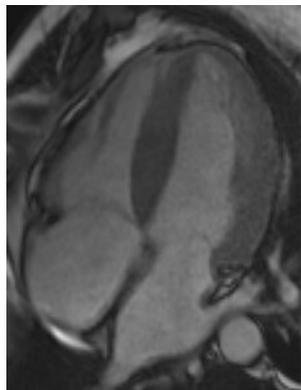
Adolescenti
Intramurale setto
Rischio elevato
Sudden Death



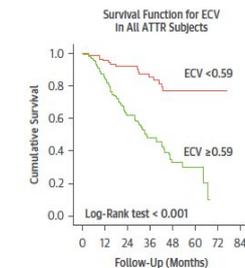
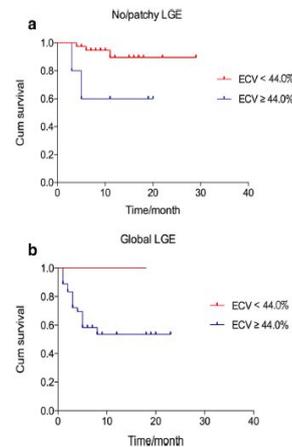


IV CASO

Amiloidosi & prognosi



T1 mapping precontrasto

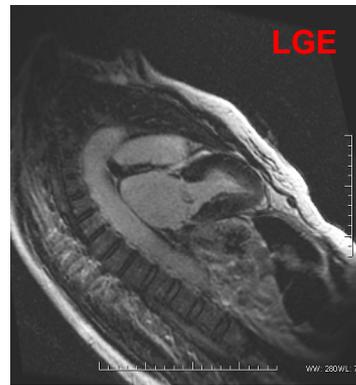
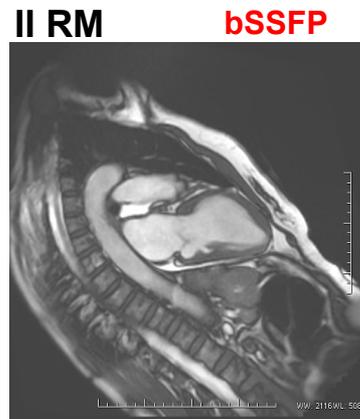
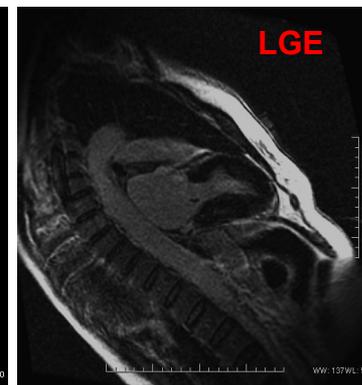


Number at risk

Follow-Up (Months)	0	12	24	36	48	60	72	84
ECV < 0.59	88	83	67	45	28	5	0	0
ECV \geq 0.59	123	105	67	38	19	4	0	0

T1 mapping postcontrasto

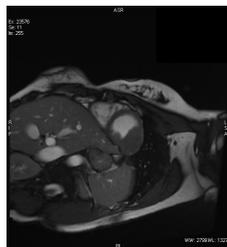
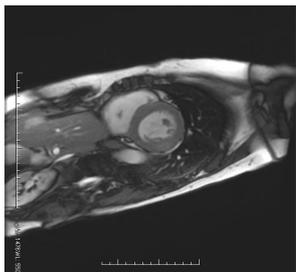
V CASO Takotsubo?





ANALISI TISSUTALE PLUS VALORE

Molte ipertrofie ventricolari sinistre per differenti malattie



European Society of Cardiology
doi:10.1093/ehjci/jex323

European Heart Journal - Cardiovascular Imaging (2017) 00, 1–10

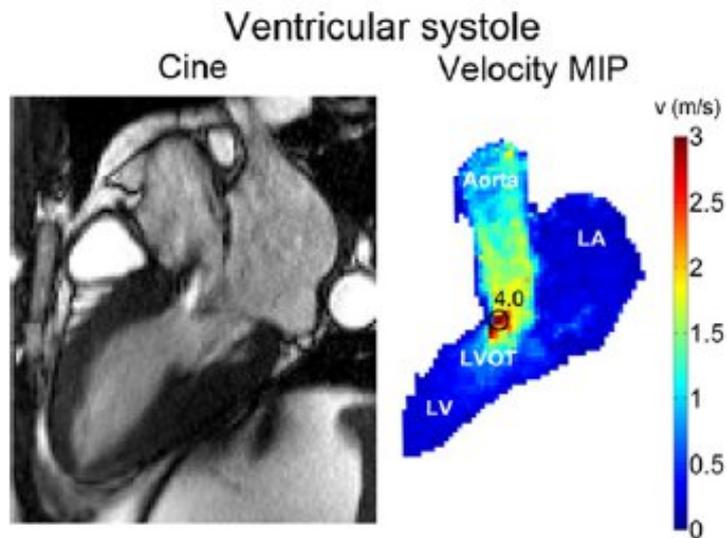
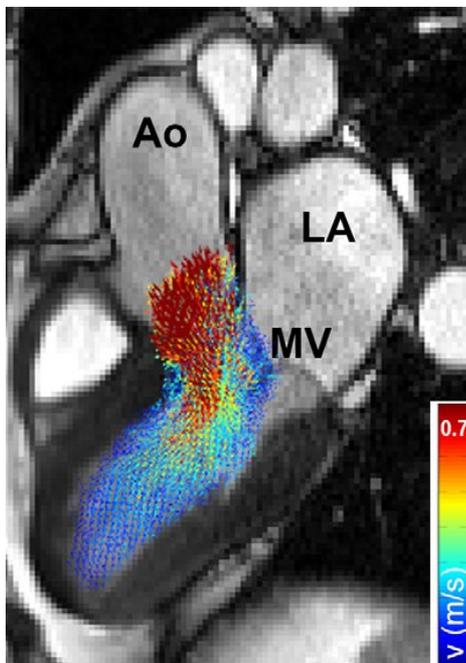
REVIEW

Cardiovascular magnetic resonance imaging in hypertrophic cardiomyopathy: the importance of clinical context

Giovanni Quarta^{1*}, Giovanni Donato Aquaro², Patrizia Pedrotti³, Gianluca Pontone⁴, Santo Dellegrottaglie⁵, Attilio Iacovoni¹, Paolo Brambilla⁶, Silvia Pradella⁷, Giancarlo Todiere², Fausto Rigo⁸, Chiara Bucciarelli-Ducci⁹, Giuseppe Limongelli¹⁰, Alberto Roghi³, and Iacopo Olivetto¹¹



4D FLOW : Future????



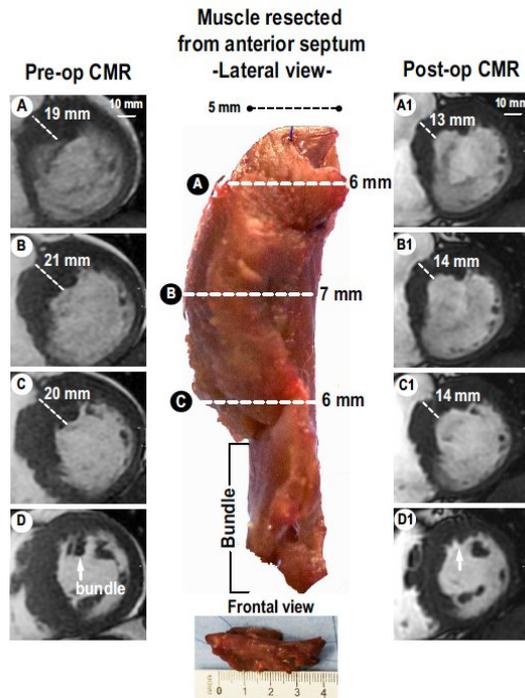
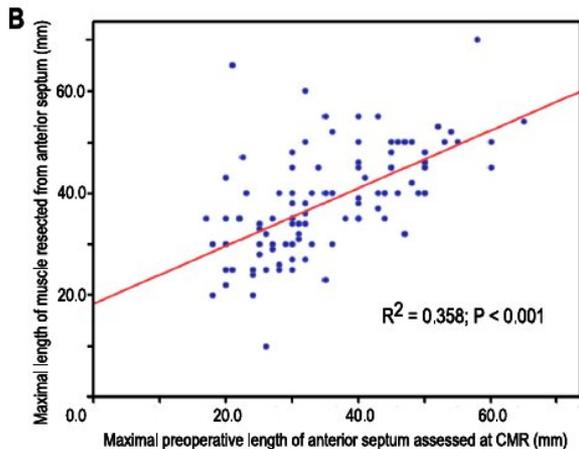
Pruijssen et al. Radiology: Cardiothoracic Imaging 2020



Role of Preoperative Cardiovascular Magnetic Resonance in Planning Ventricular Septal Myectomy in Patients With Obstructive Hypertrophic Cardiomyopathy

Paolo Spirito, MD^{1,2}, Irene Binaco, MD³, Daniele Poggio, MD⁴, Aleksei Zyrianov, MD⁵,
 Massimiliano Grillo, MD², Laura Pezzoli, PhD², Jessica Rossi, MD², Dmitri Malanin, MD²,
 Giuseppe Vaccari, MD², Lucian Dorobantu, MD^{1,2}, Maria Iacone, PhD², Andrea Mortara, MD²,
 Toufic Khouri, MD³, Paolo Bruzzi, PhD MD¹, and Paolo Ferrazzi, MD³

2019



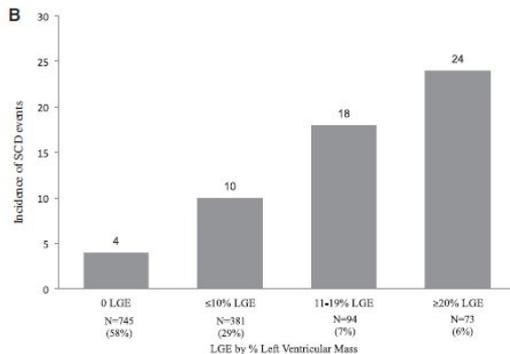
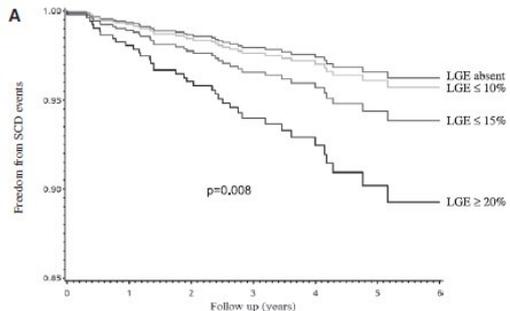
Outcome after septal myectomy in 112 patients in whom the extent of the septal excision was planned preoperatively at CMR.

Variable	Before surgery	After surgery	p value*
No. of in-hospital deaths	—	1 (0.9%)	—
No. of iatrogenic septal defect	—	0	—
NYHA functional class [†]			<0.0001
I	0	80 (72%)	
II	20 (18%)	27 (24%)	
III-IV	91 (82%)	4 (3.6%)	
Atrial fibrillation [‡]	22 (20%)	10 (9%)	<0.001
<i>Echocardiographic data</i> [‡]			
LVOT gradient at rest (mm Hg)	67 ± 41	6 ± 7	<0.0001
Post-op resting gradient ≥ 30 mm Hg (n.)	—	0	
Maximum septal thickness (mm)	23 ± 5	15 ± 3	<0.0001
Mitral valve regurgitation grade [‡]			<0.0001
0-1	43 (40%)	88 (82%)	
2	50 (48%)	17 (15%)	
3	12 (11%)	2 (2%)	
4	2 (2%)	0	
Mitral valve replacement	—	4 (3.6%)	

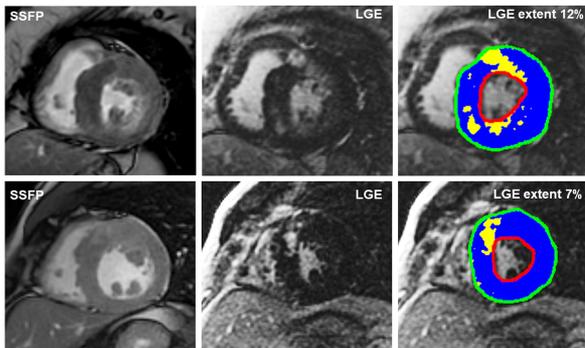


Prognostic Value of Quantitative Contrast-Enhanced Cardiovascular Magnetic Resonance for the Evaluation of Sudden Death Risk in Patients With Hypertrophic Cardiomyopathy
 Raymond H. Chan, Barry J. Maron, Jacopo Olivetto, Michael J. Pencina, Gabriele Egidio Assezzo, Tammy Haas, John R. Lesser, Christiane Gruner, Andrew M. Crean, Harry Rakowski, James E. Udelson, Ethan Rowin, Massimo Lombardi, Franco Cecchi, Benedetta Tomberli, Paolo Spirito, Francesco Fornisano, Elena Biagini, Claudio Rapezzi, Carlo Nicola De Cecco, Camillo Autore, E. Francis Cook, Susie N. Hong, C. Michael Gibson, Warren J. Manning, Evan Appelbaum and Martin S. Maron

LGE extent & prognosis («quantitative assessment»)



LGE > 15% (6 SD) della massa ventricolare sinistra: rischio di SD aumentato di 2 volte



Recommended assessment:
History
2D/Doppler Echo

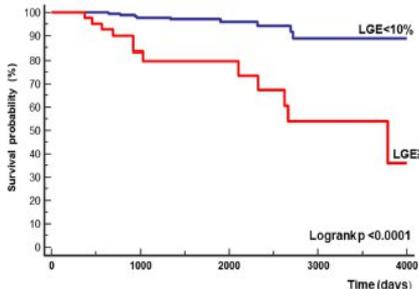
48h ambulatory ECG
HCM Risk-SCD Score

LOW RISK
 5-year risk < 4%

INTERMEDIATE RISK
 5-year risk ≥ 4-<6%

HIGH RISK
 5-year risk ≥ 6%

354 pz con ESC risk score <6%/5 anni
 Survival from Hard Cardiac Events



Number at risk	0	1000	2000	3000	4000
LGE<10%:	261	164	69	27	6
LGE≥10%:	70	22	14	3	2

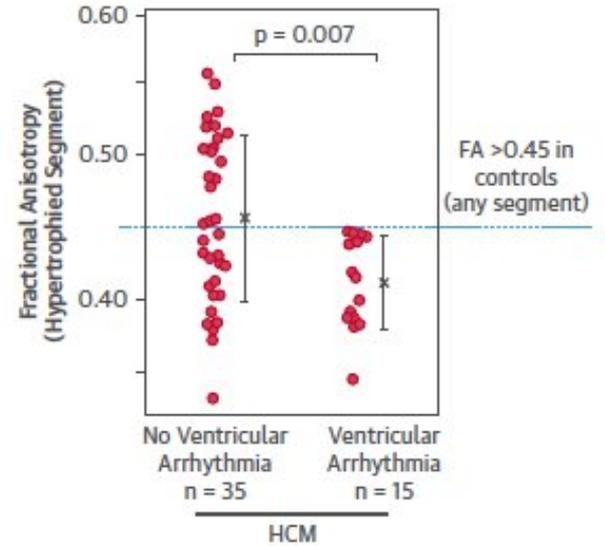
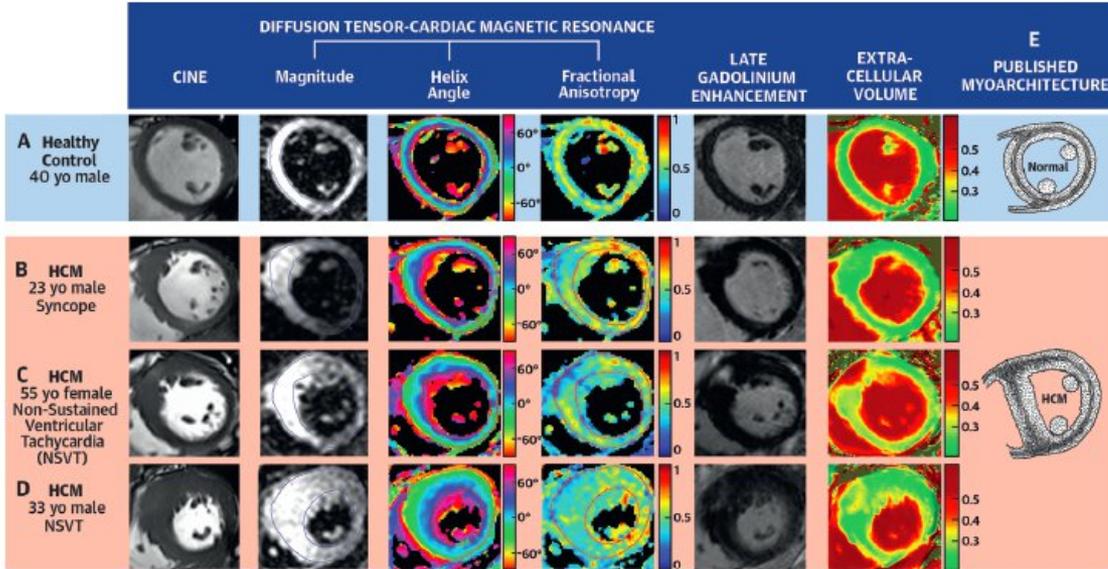
CARDIAC MAGNETIC RESONANCE
Quantitative LGE% by CMR

LGE < 10%
LOW RISK
 5-year risk 2.5%

LGE ≥ 10%
HIGH RISK
 5-year risk 22.4%

ICD generally not indicated

ICD should be considered



Ariga et al. JACC 2019



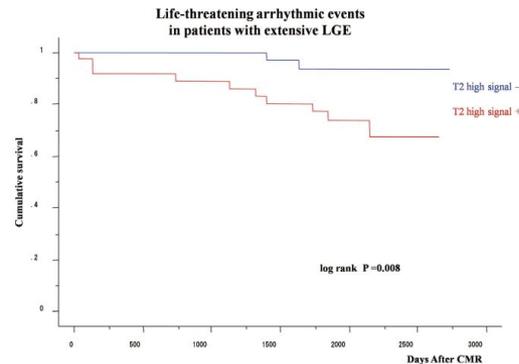
Abnormal T2-STIR Magnetic Resonance in Hypertrophic Cardiomyopathy: A Marker of Advanced Disease and Electrical Myocardial Instability

Giancarlo Todiere^{1*}, Lorena Pisciella², Andrea Barison¹, Annamaria Del Franco¹, Elisabetta Zachara², Paolo Piaggi³, Federica Re², Alessandro Pingitore⁴, Michele Emdin¹, Massimo Lombardi⁵, Giovanni Donato Aquaro^{1*}

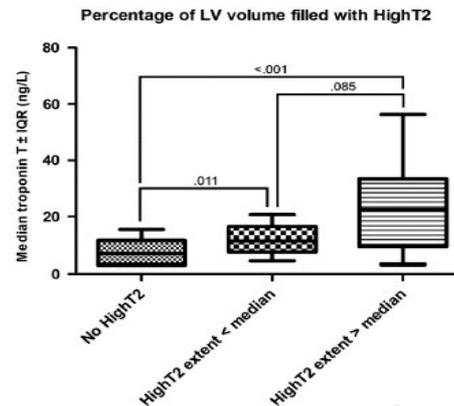


Independent variables	Logistic regression Analysis						
	NSVT	No NSVT	p value	Coefficient	Odds Ratio	95%-C.I.	p-value
Arrhythmic Risk factors	0.6±0.7	0.14±0.3	0.001				
LV mass index	131±38	103±39	0.007				
HyT2	21(95)	1(5)	<0.001	5.1	165	11-2455	<0.001
Maximal Wall Thickness	25±6	19±6	<0.001	0.12	1.1	1.0-1.3	<0.001
Extent of LGE	18.3±11.9	6.8±7.9	<0.001				

PlosONE 2014

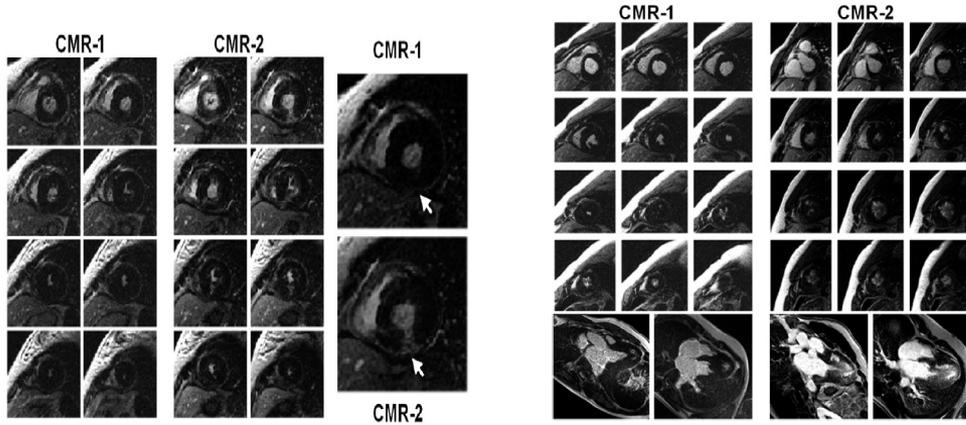


Hen y et al Circ J 2018



Danno miocardico

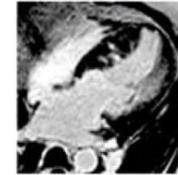
Gommans DHF Heart 2016



Todiere G, Aquaro GD JACC 2012

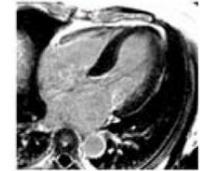
2755 HCM patients
44 sites
6 countries
North America and Europe

2 broad, relatively distinct populations



Sarcomere mutation (+)
More Likely:

Reverse septal curvature morphology
More LGE and interstitial fibrosis
No significant LVOT obstruction



Sarcomere mutation (-)
More Likely:

Isolated basal septal morphology
Less LGE and interstitial fibrosis
More LVOT obstruction

Neubauer S et al JACC 2019



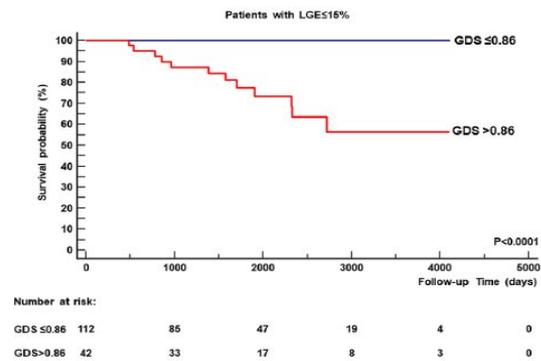
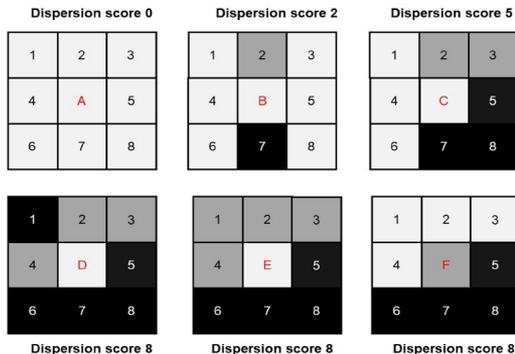
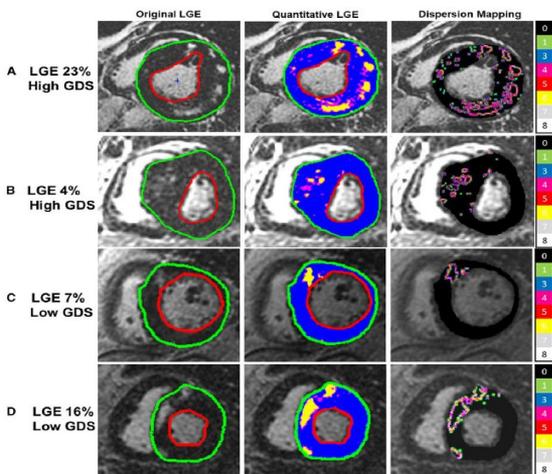


Circulation: Cardiovascular Imaging

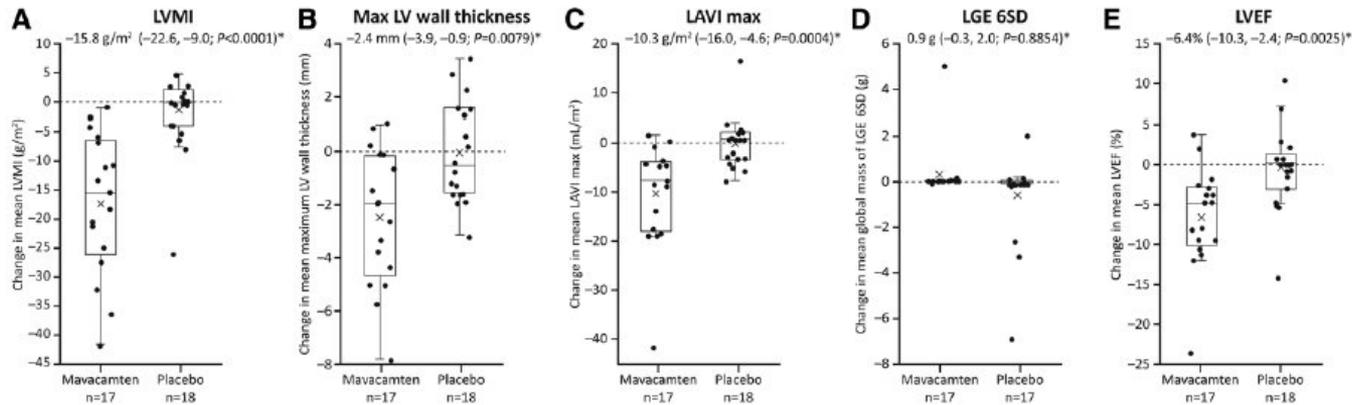
ORIGINAL ARTICLE

Late Gadolinium Enhancement–Dispersion Mapping

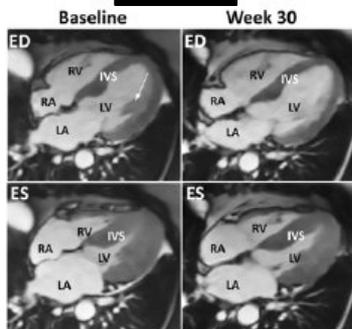
A New Magnetic Resonance Imaging Technique to Assess Prognosis in Patients With Hypertrophic Cardiomyopathy and Low-Intermediate 5-Year Risk of Sudden Death



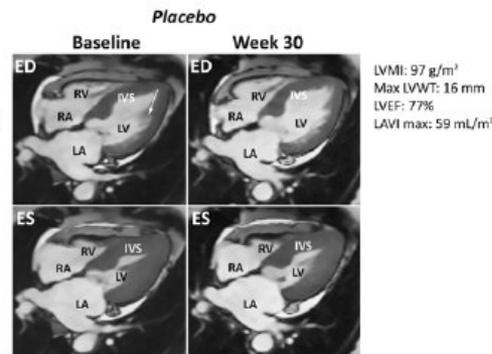
Aquaro GD et al. 2021



F
 CAMZYOS



G
 PLACEBO





TAKE HOME MESSAGES

RM ruolo diagnostico imprescindibile a patto che sia eseguita al massimo delle sue potenzialità

Contesto clinico rimane comunque fondamentale...

Possibilità di ripetere la RM nel tempo potendo monitorare «fasi di attività» della malattia, e la progressione delle alterazioni strutturali della cardiomiopatia nel tempo