Ipertrofia segno di salute o di malattia?

Il sottile discrimine tra ipertrofia dell'Atleta e Cardiomiopatia Ipertrofica. Quando è necessaria la RM per fare la diagnosi

Giancarlo Casolo

Premesse

- Ipertrofia: aumento della massa o aumento degli spessori?
- Il Cuore d'Atleta
- Diagnosi di cardiomiopatia ipertrofica con Risonanza Magnetica
- Ambiti clinici di possibile utilità della RM nella diagnosi differenziale

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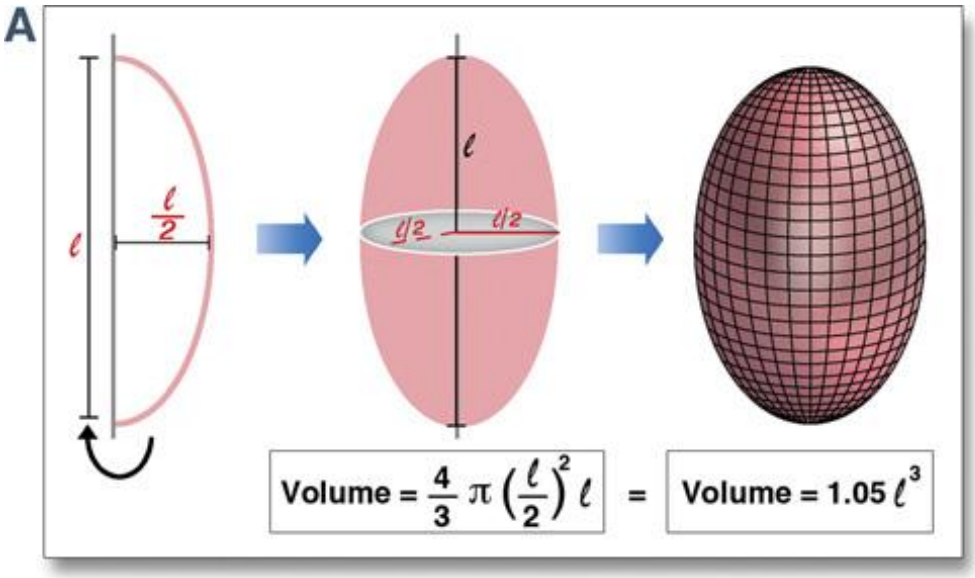
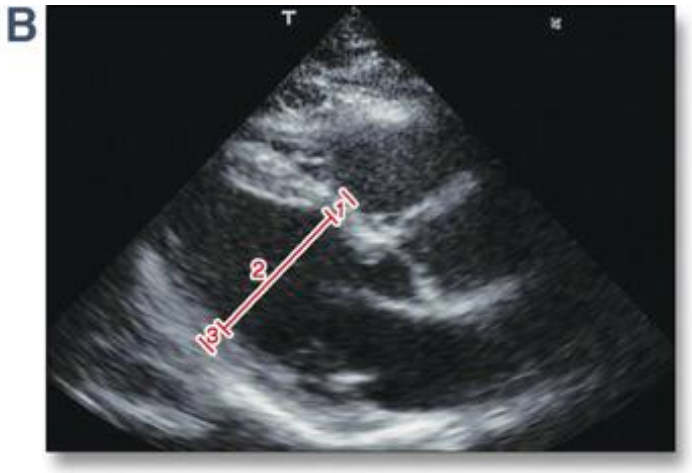
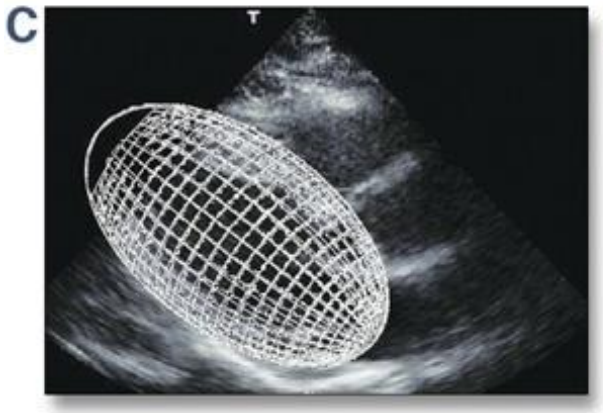






$\text{Volume}_T = 1.05\left(\text{IVST}+\text{LVID}+\text{PWT}\right)^3$

$\text{Volume}_C = 1.05\left(\text{LVID}\right)^3$

$\text{Volume}_M = \text{Volume}_T - \text{Volume}_C$

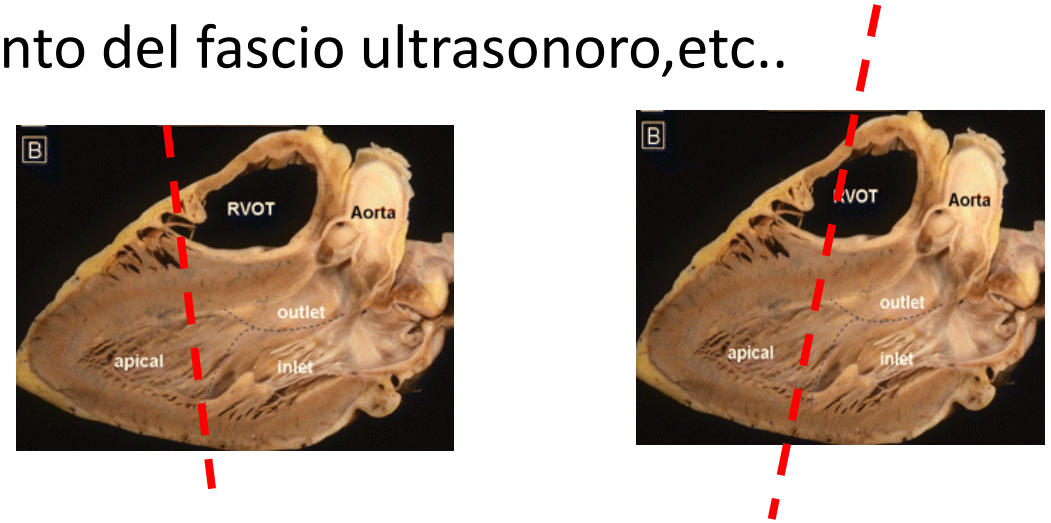
F

$$\text{LVM} + 0.8 \times \left[1.05\left(\text{IVST}+\text{LVID}+\text{PWT}\right)^3 - \left(\text{LVID}\right)^3\right] + 0.8\ \text{g}$$

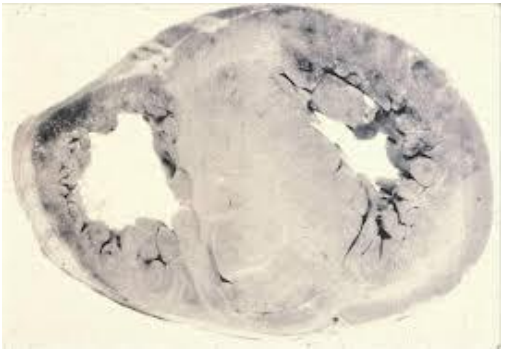
Armstrong et Al. *J Am Coll Cardiol Img*. 2012

Limiti della misurazione della massa con ecocardiografia

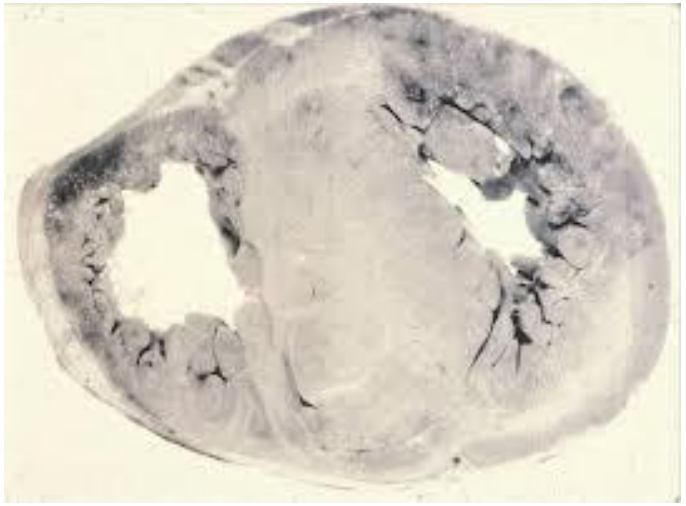
- Aspetti tecnici
  - Finestra, allineamento del fascio ultrasonoro,etc..

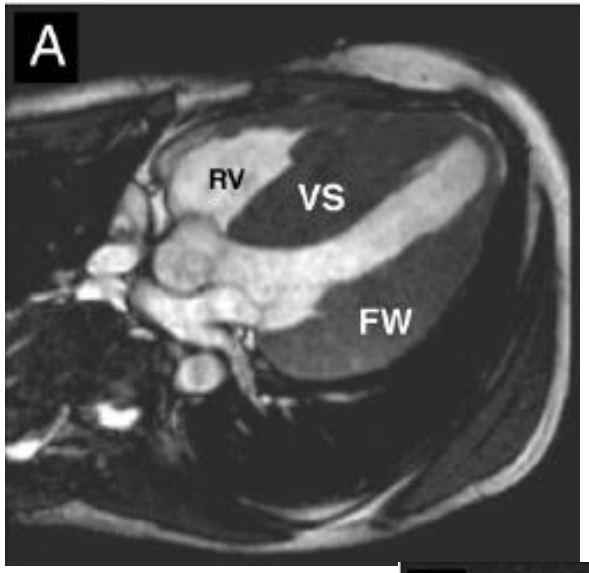


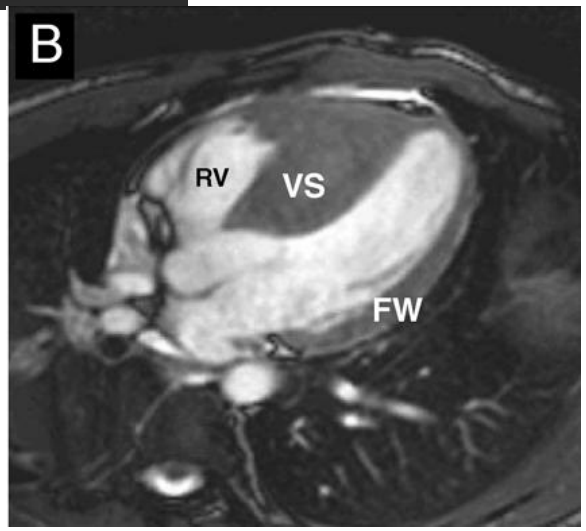
- Geometria del cuore

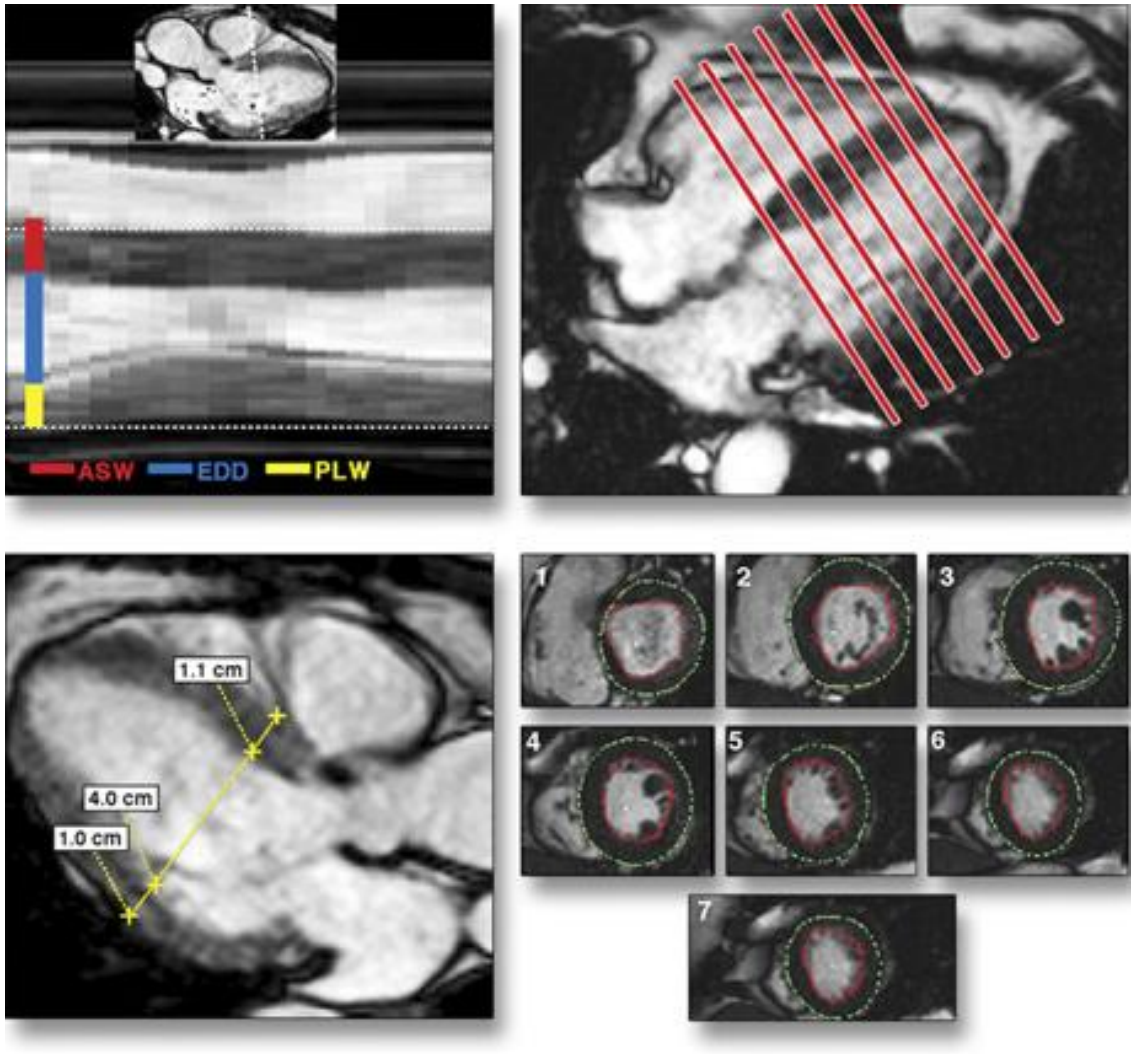


Limiti della misurazione della massa con stime geometriche



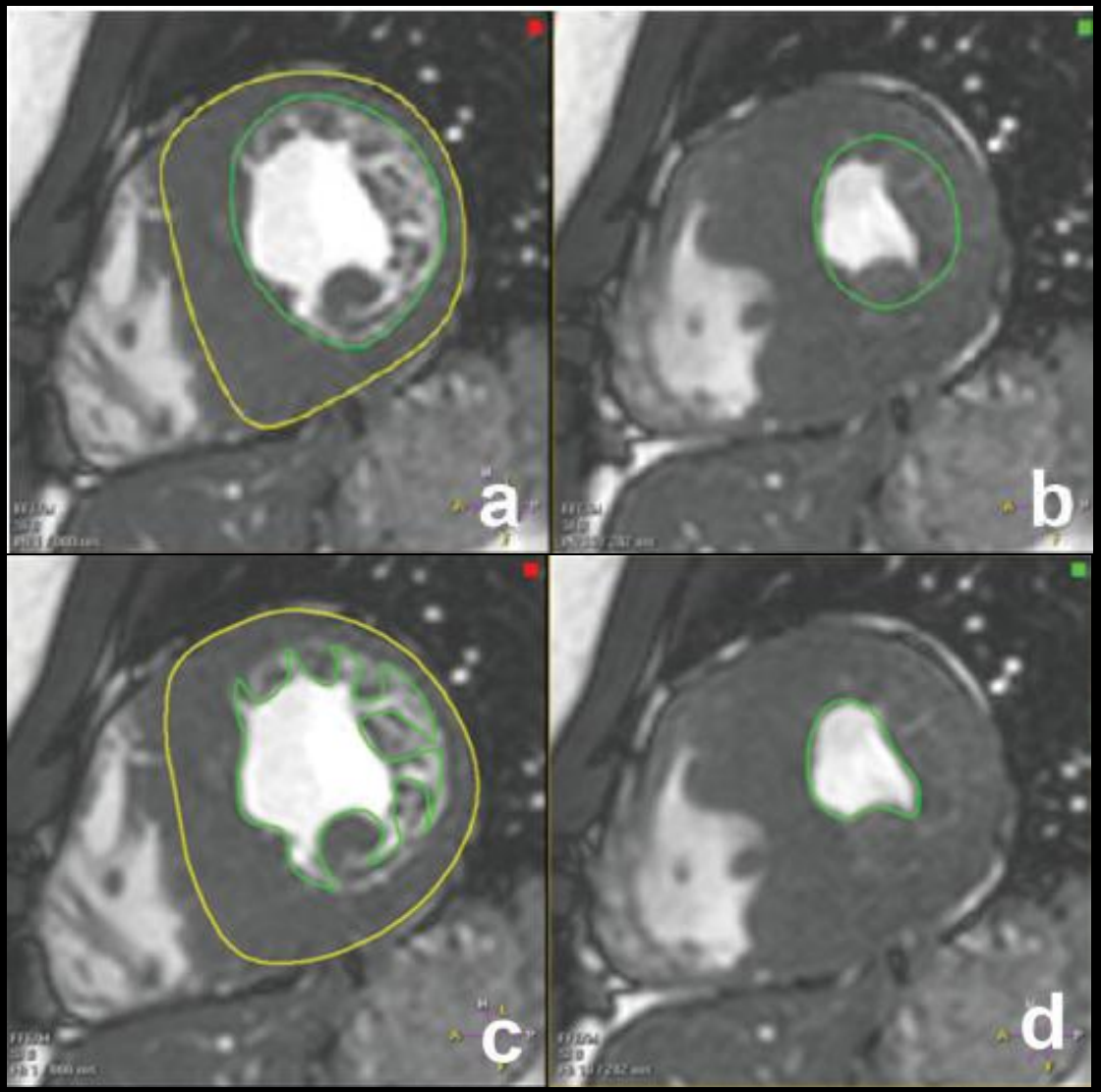




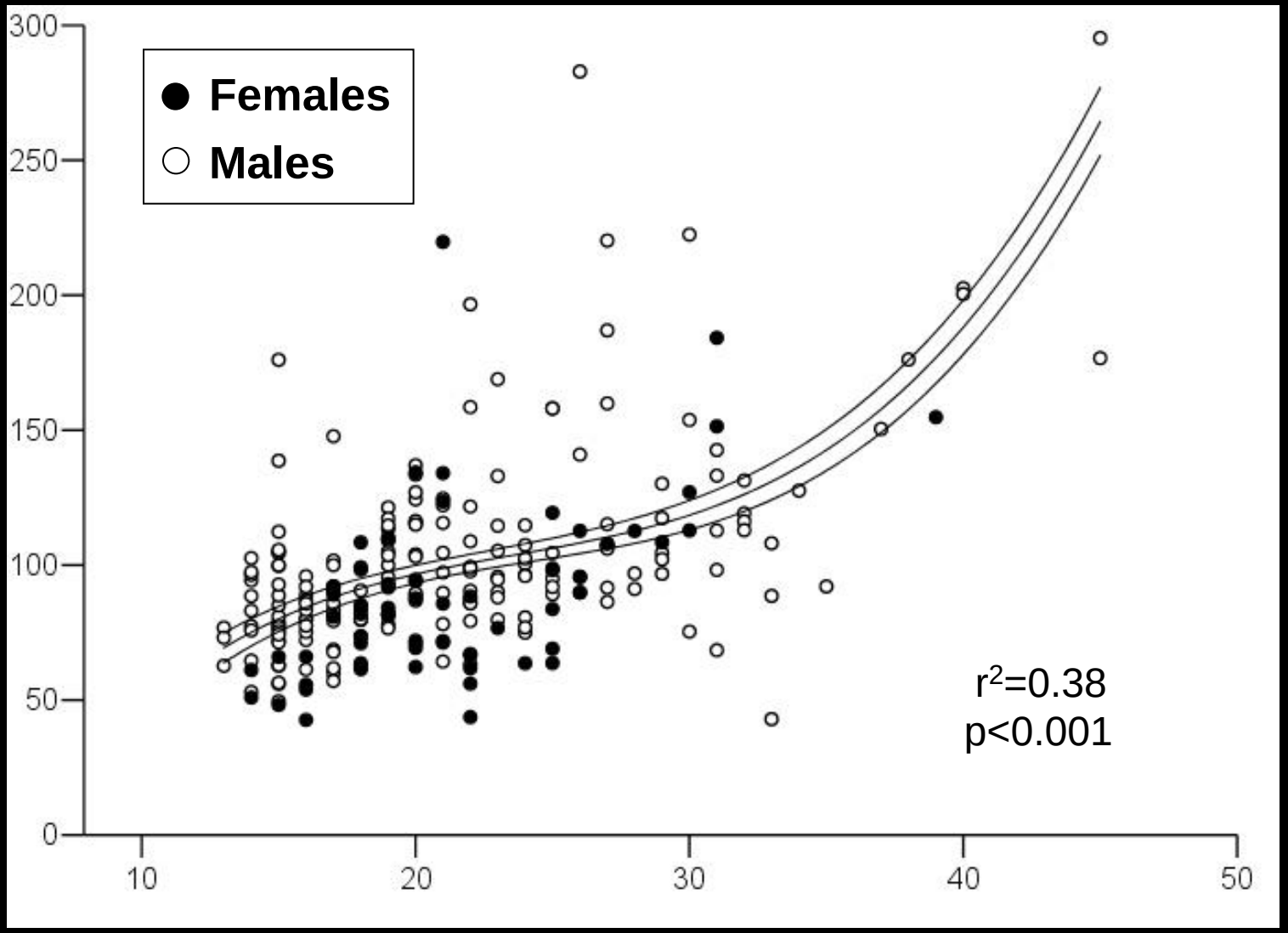


Armstrong et Al. *J Am Coll Cardiol Img*. 2012

Contouring Methods



Han et Al. J. Magn. Reson. Imaging 2009



Olivotto et Al. JACC 2008

Premesse

- Ipertrofia: aumento della massa o aumento degli spessori?
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- Diagnosi di cardiomiopatia ipertrofica con Risonanza Magnetica
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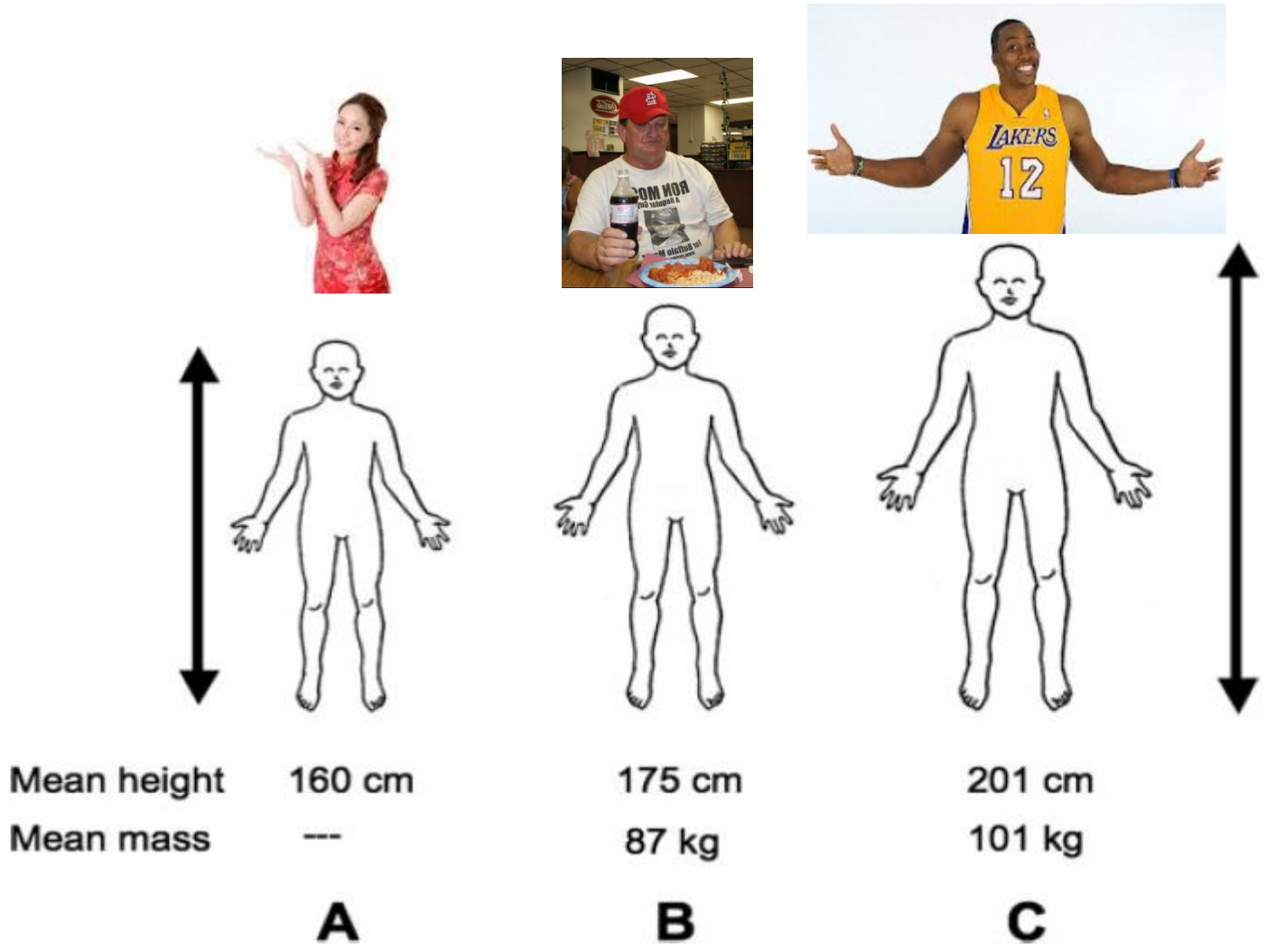
Does Size Matter? Clinical Applications of Scaling Cardiac Size and Function for Body Size



Dewey et Al. Circulation 2008

Circulation
Cardiovascular Imaging

American Heart Association®

Circ Cardiovasc Imaging. 2013;6:1073-1079

Ventricular Structure and Function

Size Matters! Impact of Age, Sex, Height, and Weight on the Normal Heart Size

Stefan Pfaffenberger, MD; Philipp Bartko, MD; Alexandra Graf, PhD; Elisabeth Pernicka, PhD; Jamil Babayev, MD; Emina Lolic, MD; Diana Bonderman, MD; Helmut Baumgartner, MD; Gerald Maurer, MD; Julia Mascherbauer, MD

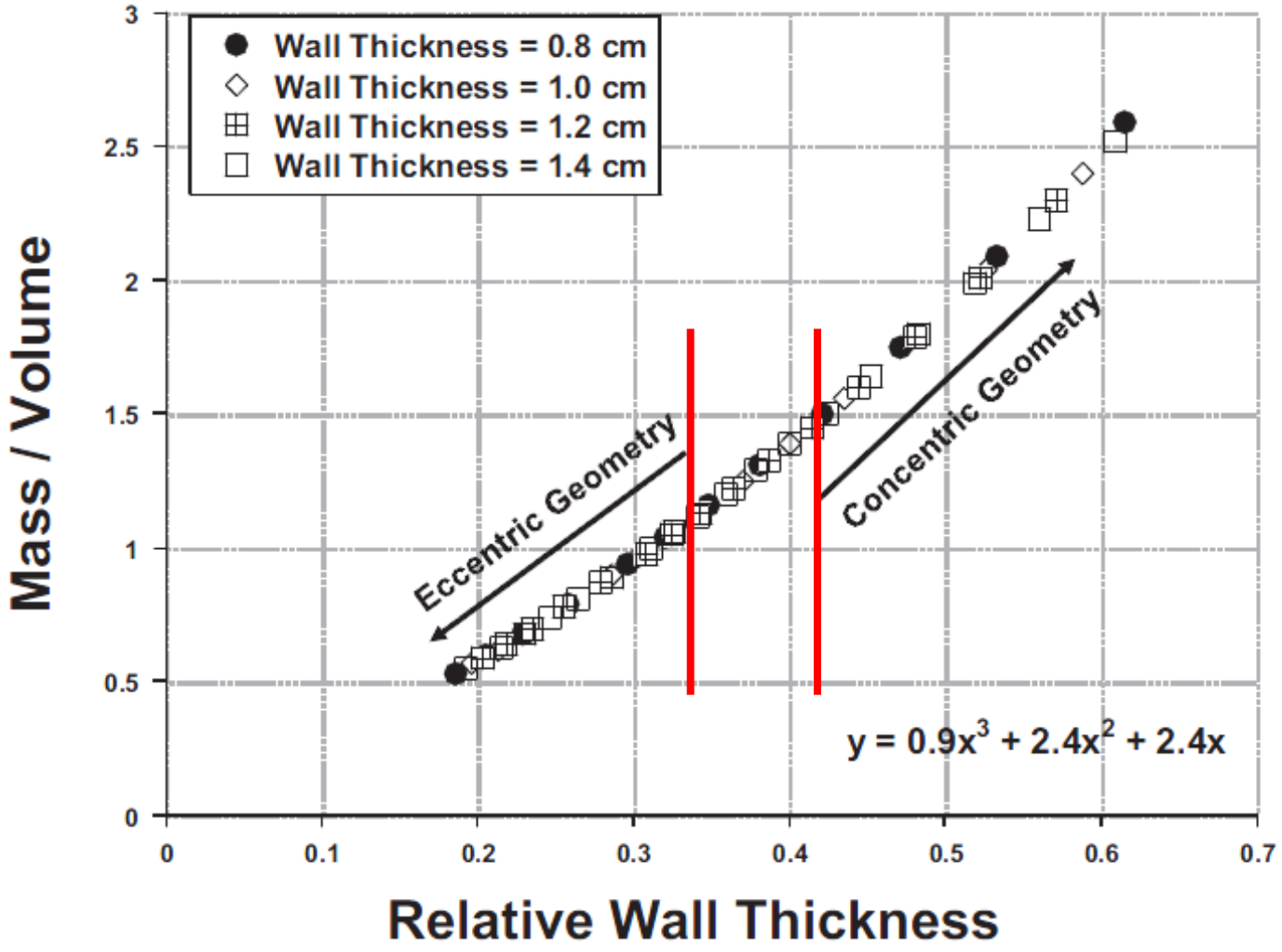
Normal Heart Size Calculator

	Height (cm)	Weight (kg)	Age (years)	Gender (male=0, female=1)	BMI (kg/m²)	BSA (m²)
Input fields →	166	85	71	1	30,8	1,98

Parameter	LV_{EDD-2D}	LV_{EDD-MM}	$LV_{EDD-Vol}$	RV_{EDD-2D}	RV_{Area}	LA_{Diam}	LA_{Area}	LA_{Vol}	RA_{Diam}	RA_{Area}	RA_{Vol}	IVS_{2D}	IVS_{MM}	PW_{MM}
intercept	20,38637	15,38196	-39,37731	15,34580	-0,37314	34,23014	-4,07280	-34,73810	35,52012	9,15336	18,37312	7,66249	7,34011	7,49088
height	0,11423	0,15007	0,73703	0,07014	0,1004	0	0,08135	0,31554	0	0	0	0	0	0
gender	-1,45756	0	-9,47838	-2,35620	-2,62433	-1,73000	0	0	-2,03817	-1,85318	-8,55635	-0,56512	-0,53678	-0,57818
age	-0,04141	0	-0,33835	0	-0,03906	0,03453	0,02773	0,10538	0,08743	0	0	0,02344	0,02463	0,01483
weight	0,07404	0,07658	0,42808	0,04375	0,03270	0,11356	0,05673	0,21533	0,10712	0,07818	0,27393	0,02031	0,01872	0,01964
Result:	**41,9**	**46,8**	**85,8**	**29,0**	**18,2**	**48,9**	**16,2**	**43,4**	**48,8**	**13,9**	**34,2**	**10,5**	**10,1**	**9,6**
Unit:	**mm**	**mm**	**ml**	**mm**	**cm²**	**mm**	**cm²**	**ml**	**mm**	**cm²**	**ml**	**mm**	**mm**	**mm**

BMI indicates body mass index; BSA, body surface area; LVEDD, left ventricular end-diastolic diameter; 2D, two-dimensional; MM, motion mode; Vol, Volume; RVEDD, right ventricular end-diastolic diameter; LA, left atrium; Diam, diameter; RA, right atrium; IVS, interventricular septum; and PW, posterior wall.

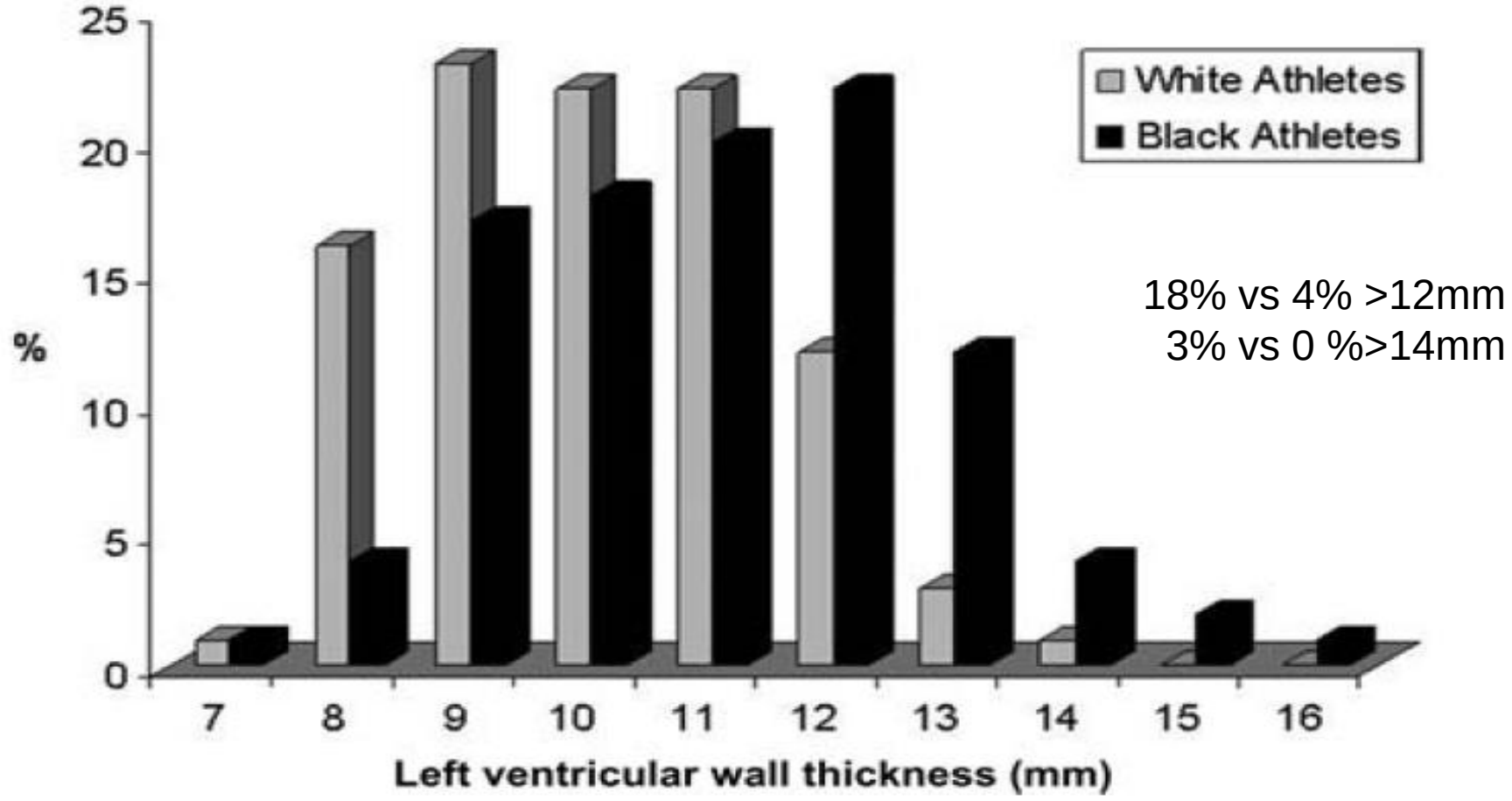
Rapporto tra spessori , volume e tipo di ipertrofia



Gaash et Al. JACC 2011

ratio of LV wall thickness to chamber radius

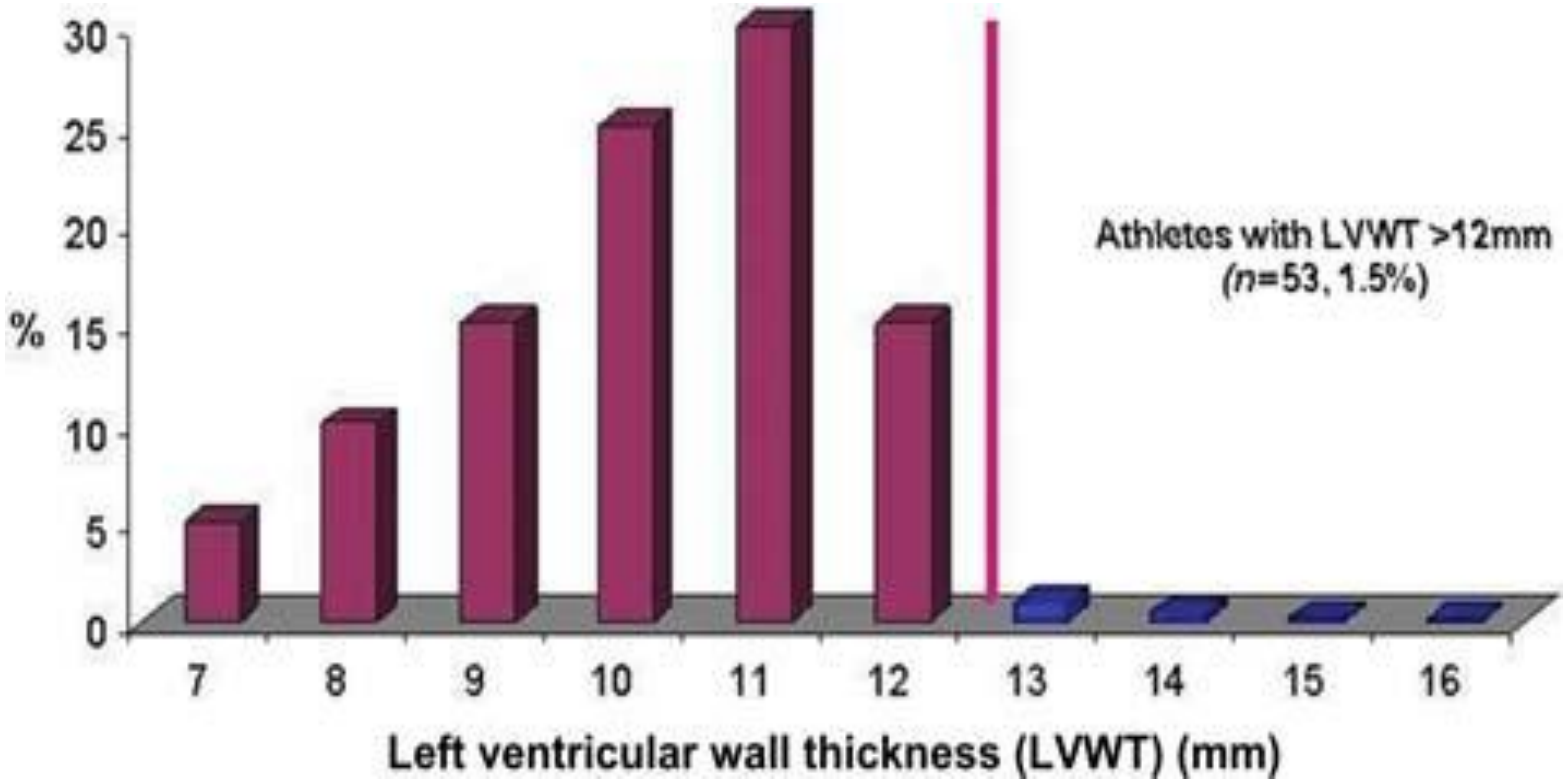
Spessori in relazione all'Etnia



Basavarajaiah et al. J Am Coll Cardiol 2008

Left ventricular hypertrophy in athletes

3500 highly trained athletes :
1.5% exhibit a LVWT >12 mm



Basavarajaiah et al. J Am Coll Cardiol 2008

Elementi definiti

- Gli atleti con spessore >12mm sono pochi, generalmente maschi e > 16 anni
- Nelle donne l'ipertrofia è assai rara, forse per il diverso profilo ormonale
- Quando presente l'ipertrofia è associata sport fortemente isometrici e/o isotonici (canoa, ciclismo, corsa di resistenza, nuoto)
- Raramente i sollevatori di peso hanno spessori >12mm
- Una superficie corporea maggiore di 2 aumenta la probabilità di osservare un aumento degli spessori

Premesse

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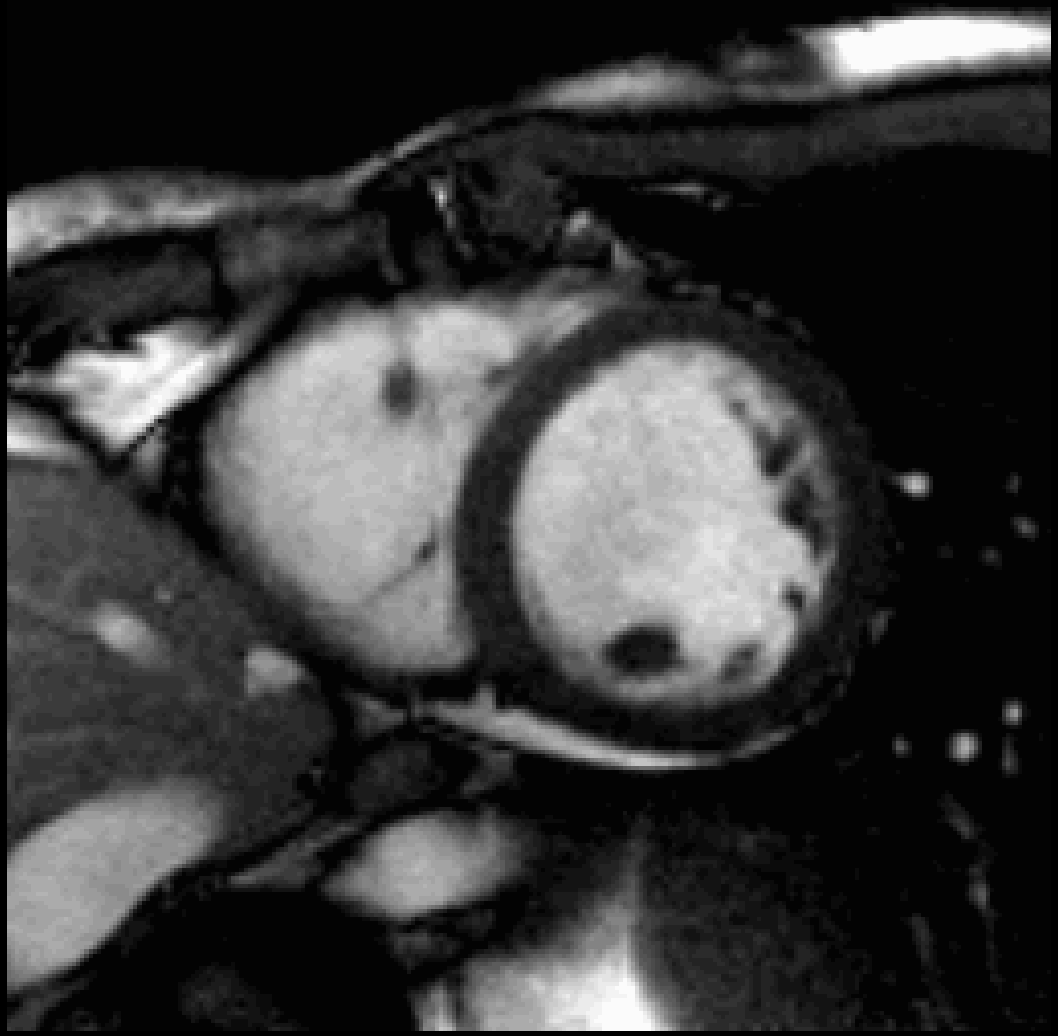
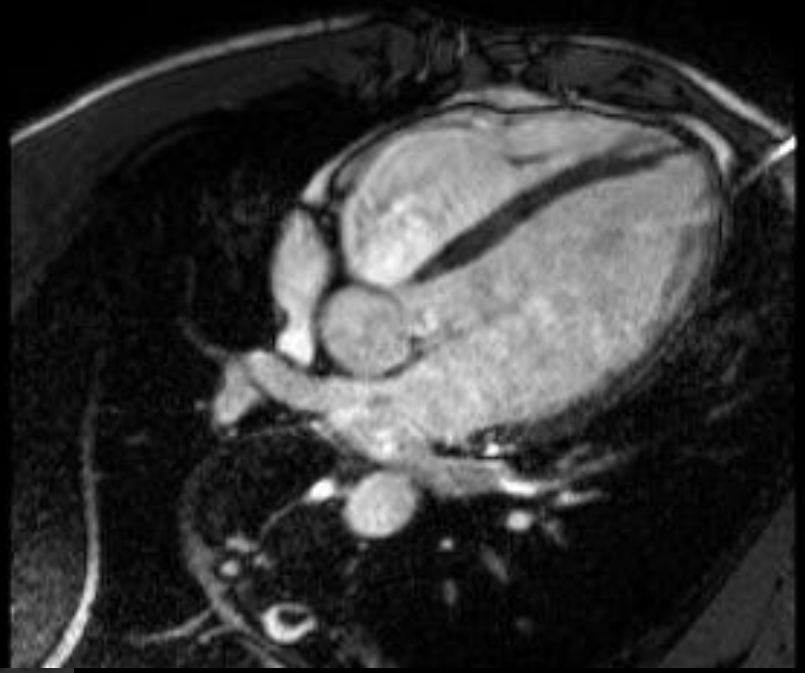
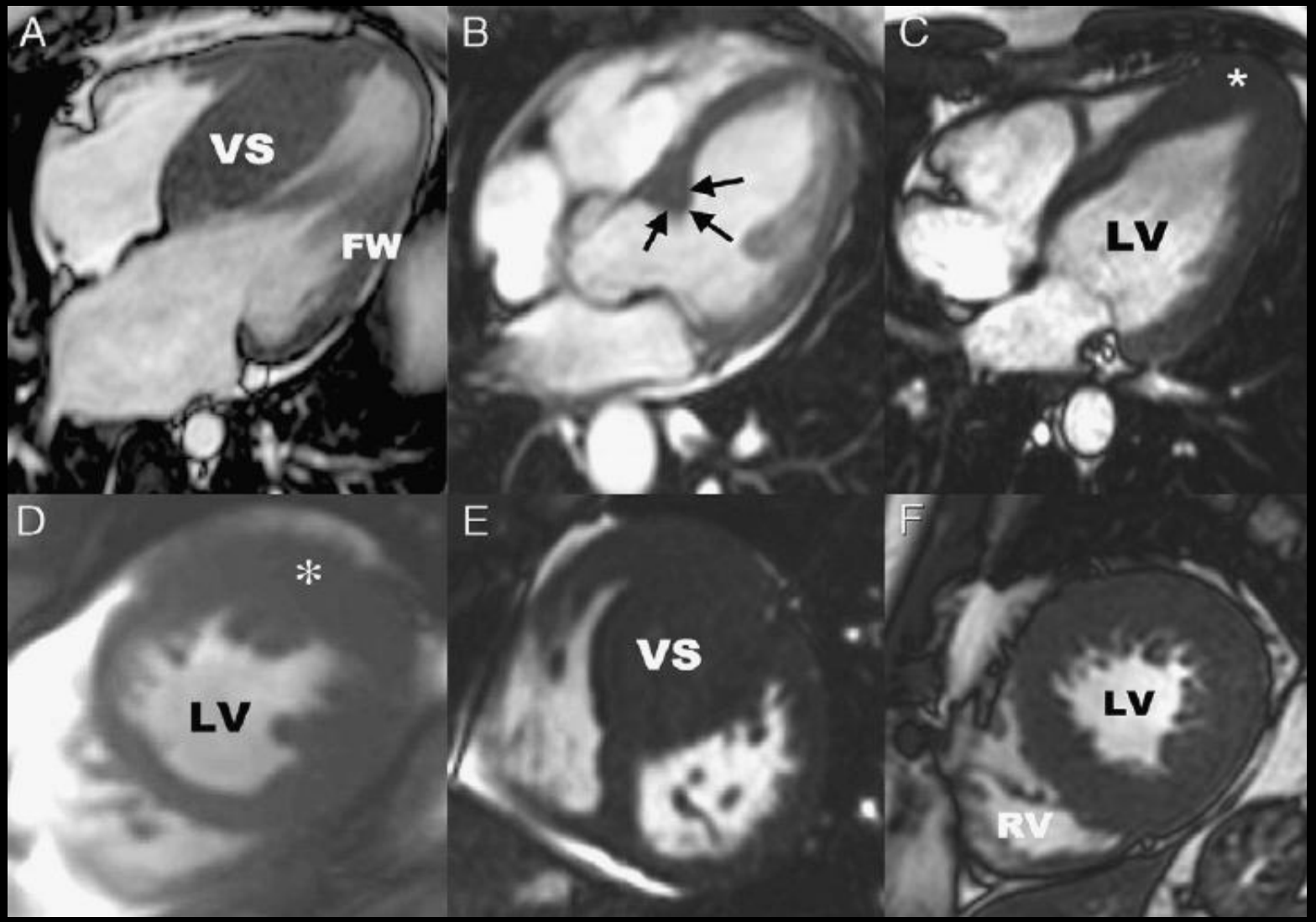


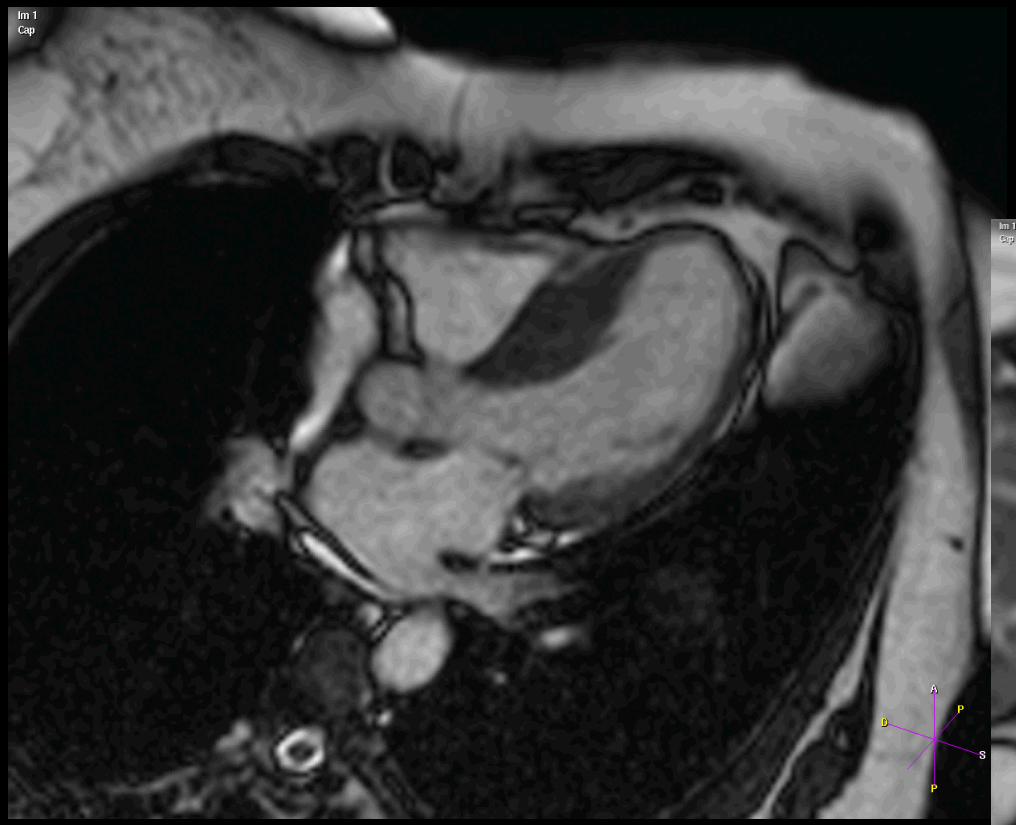


Hypertrophic Cardiomyopathy Phenotype Revisited After 50 Years With CMR



Maron et Al. JACC 2009


Im 1
Cap
A
P
D
S
P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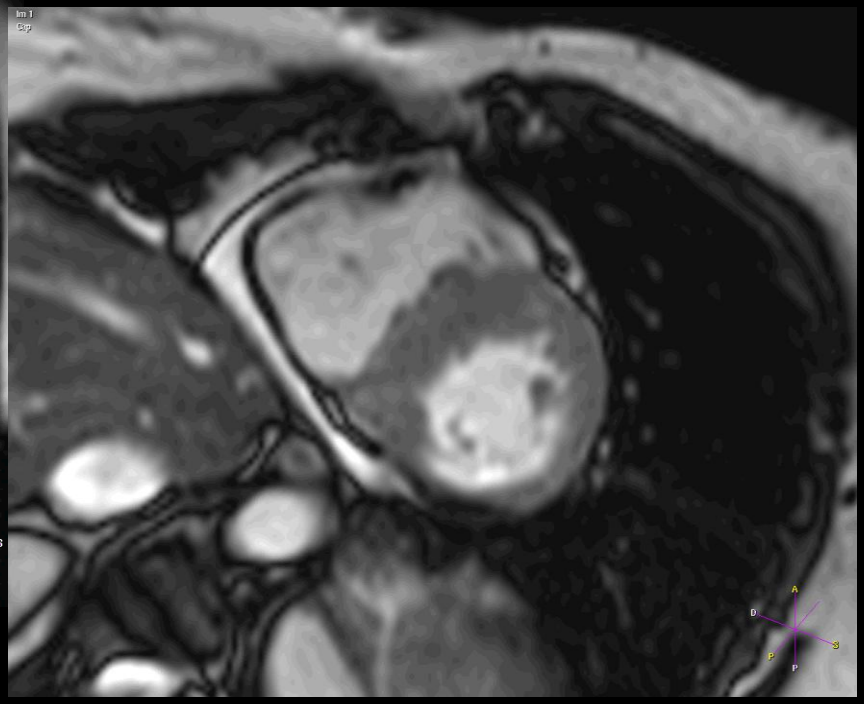
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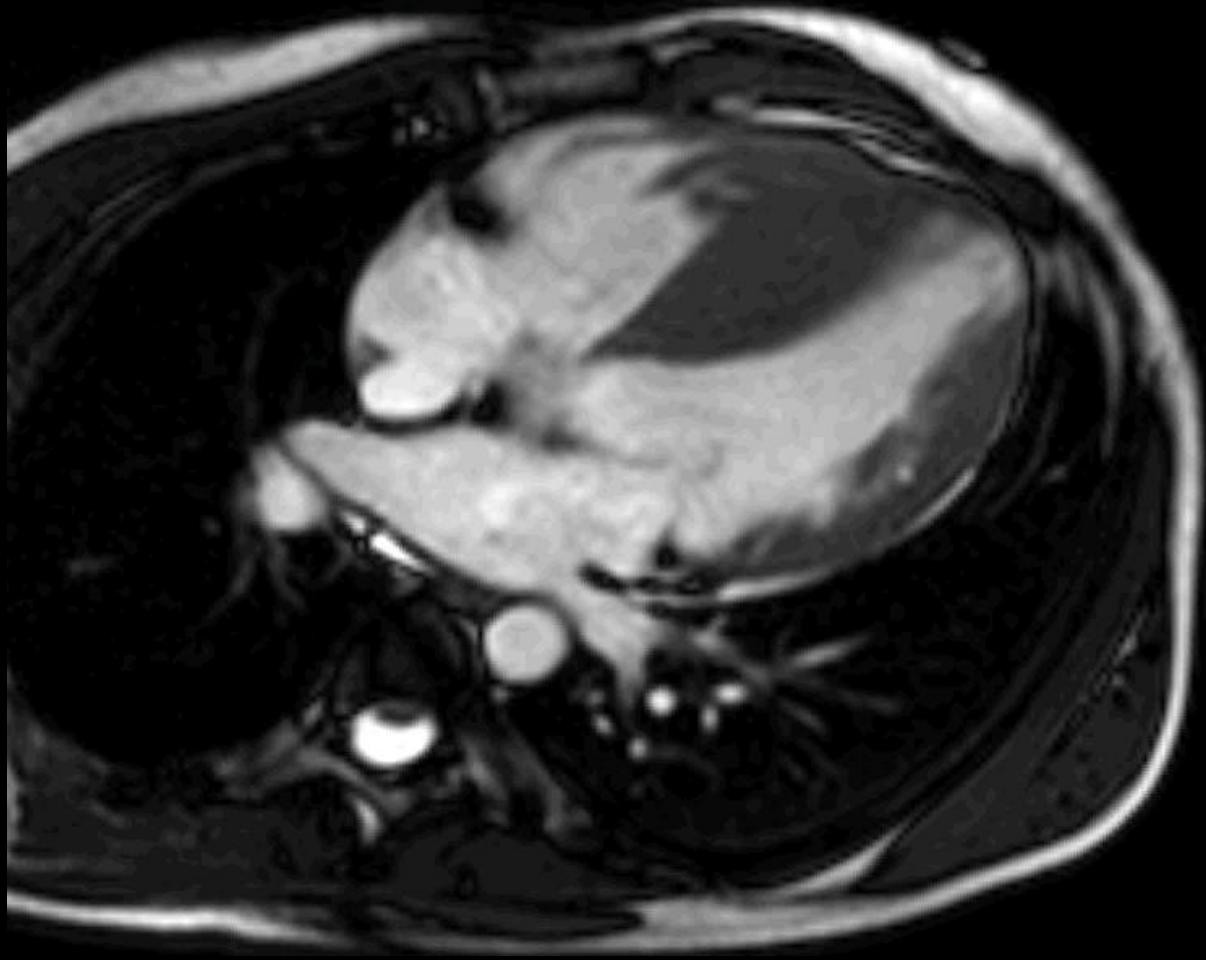

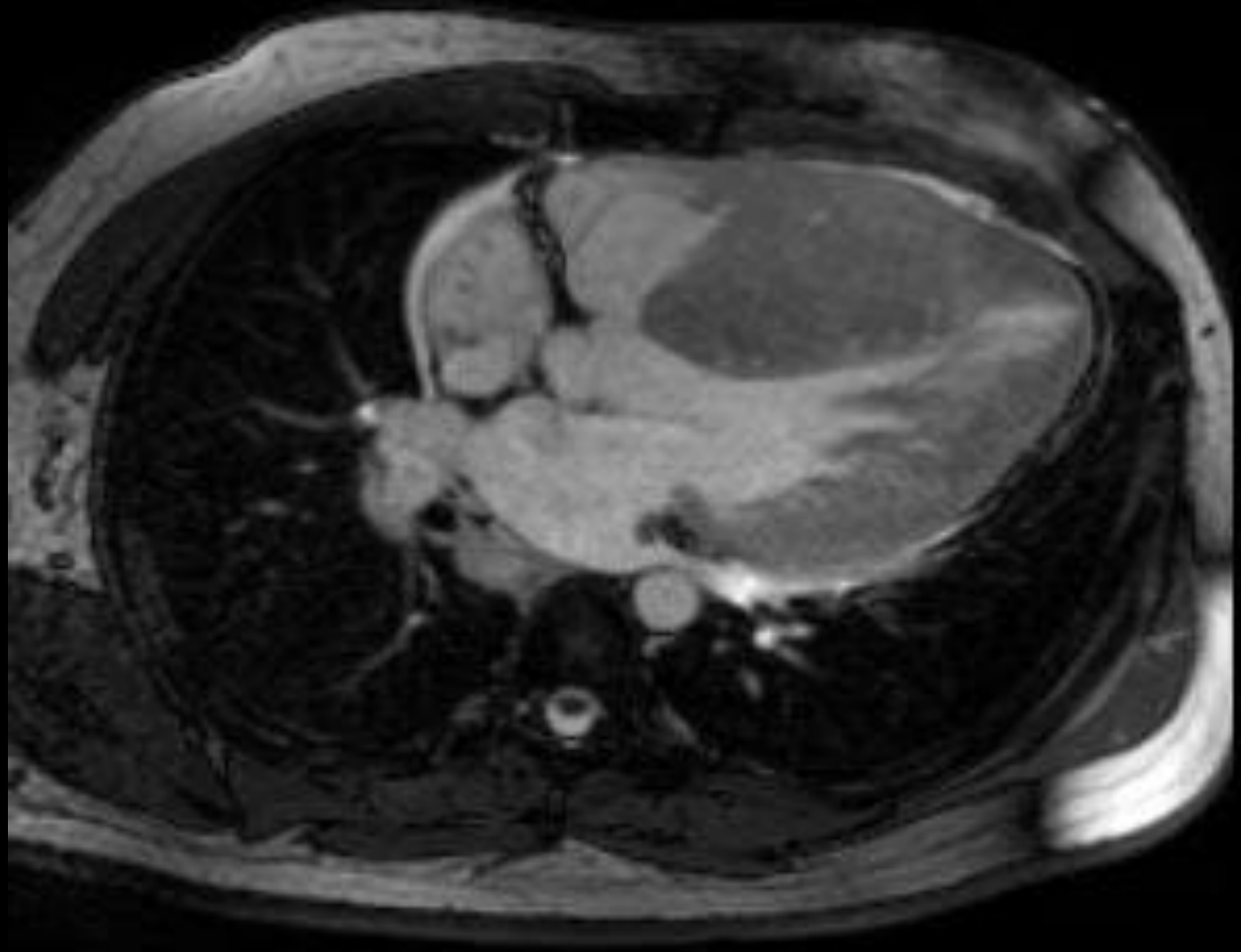

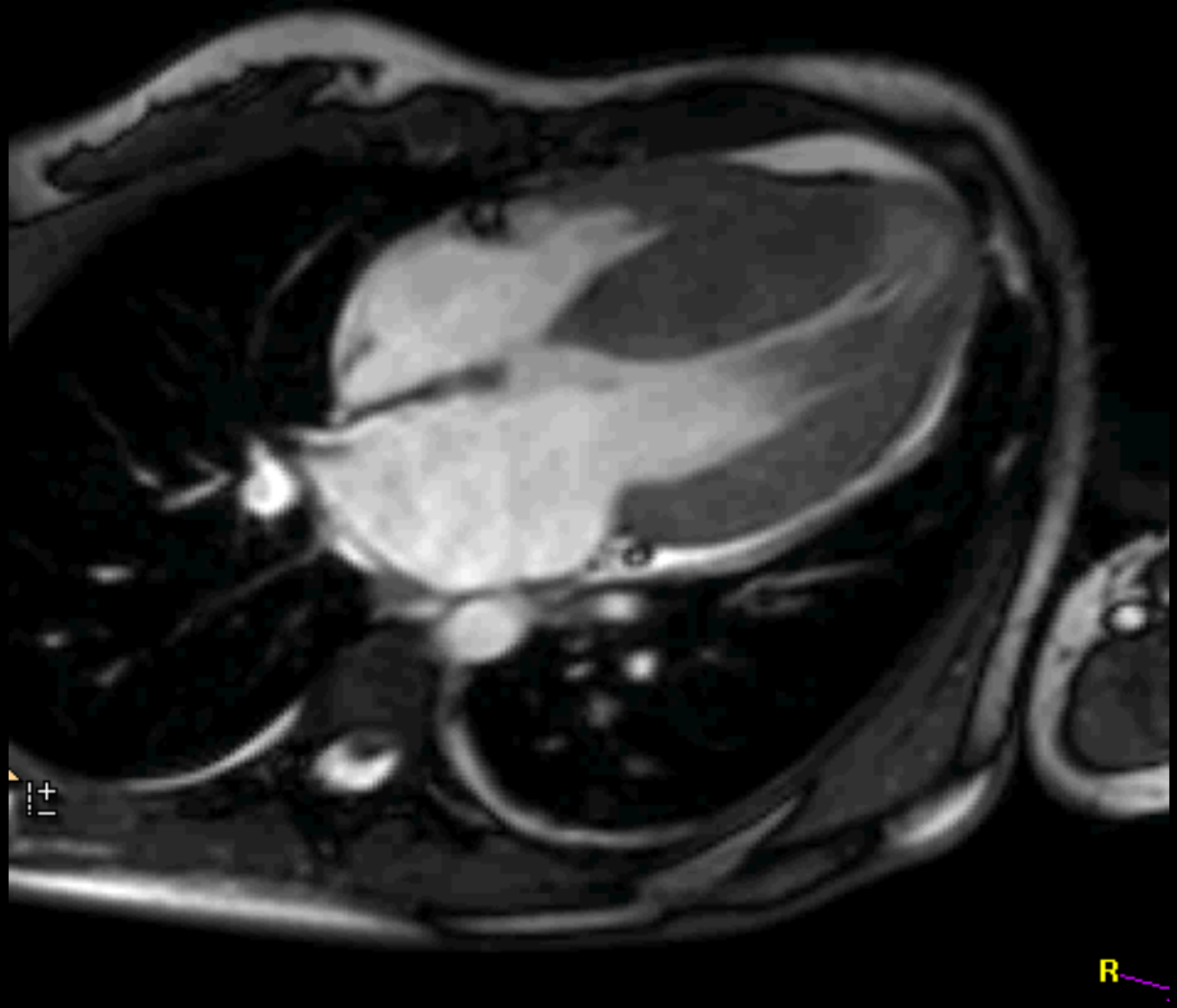
R

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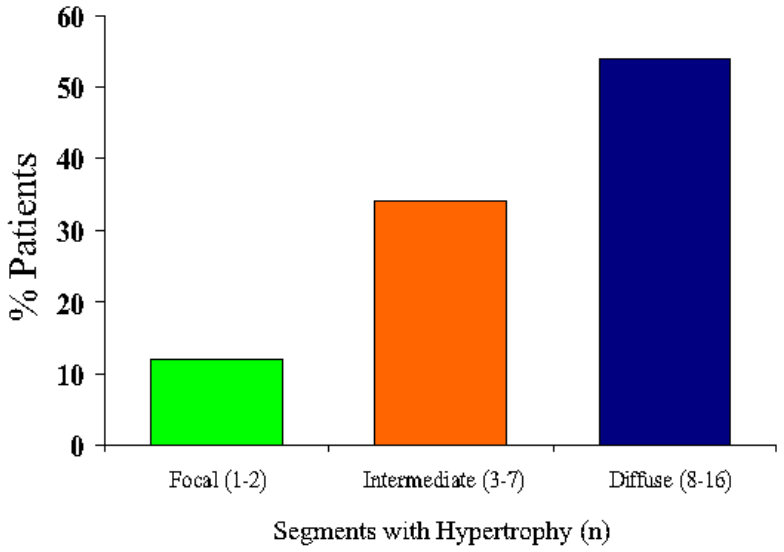
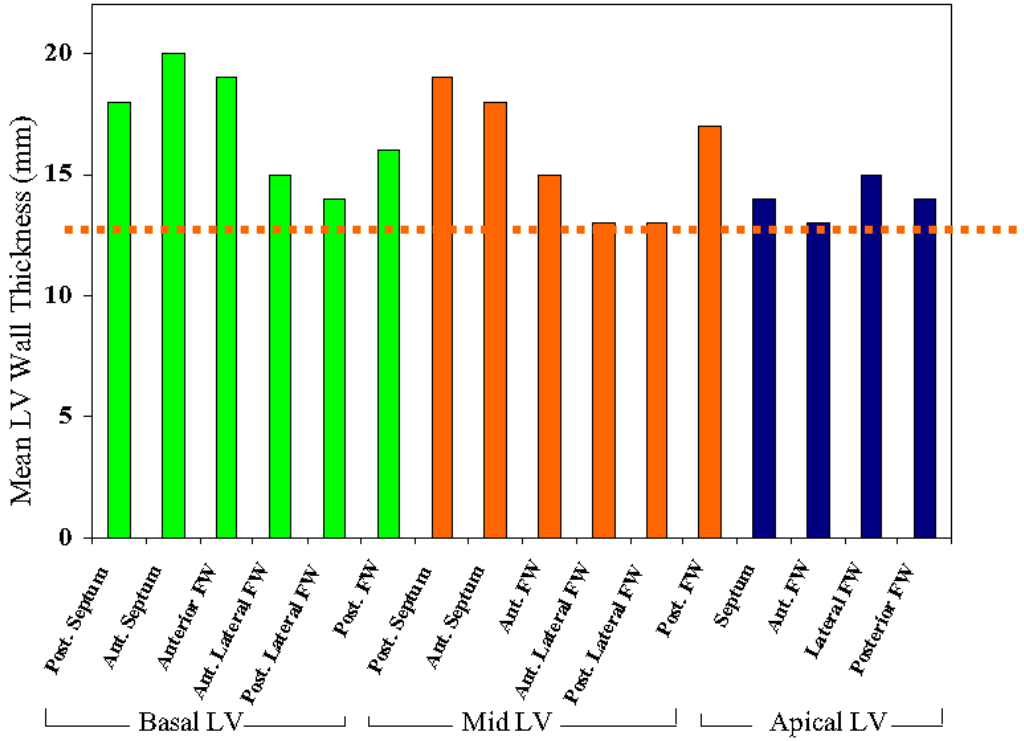
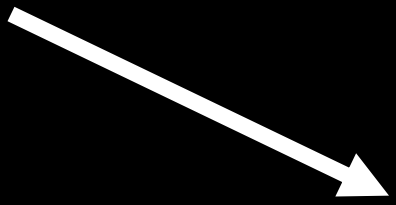


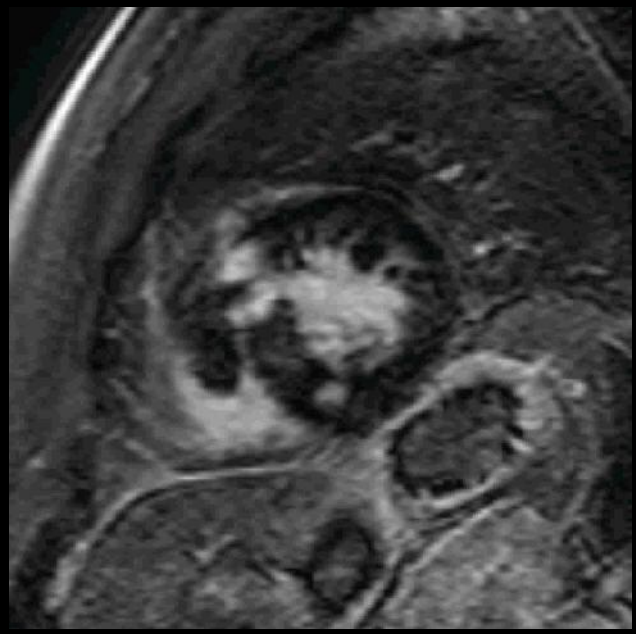
256 Pts

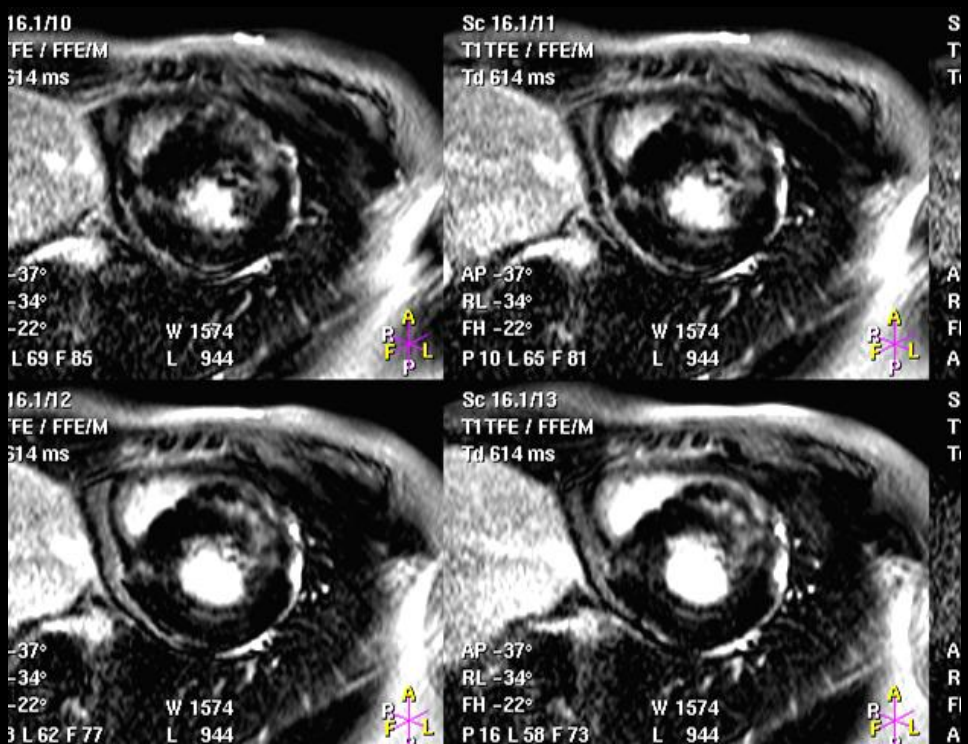


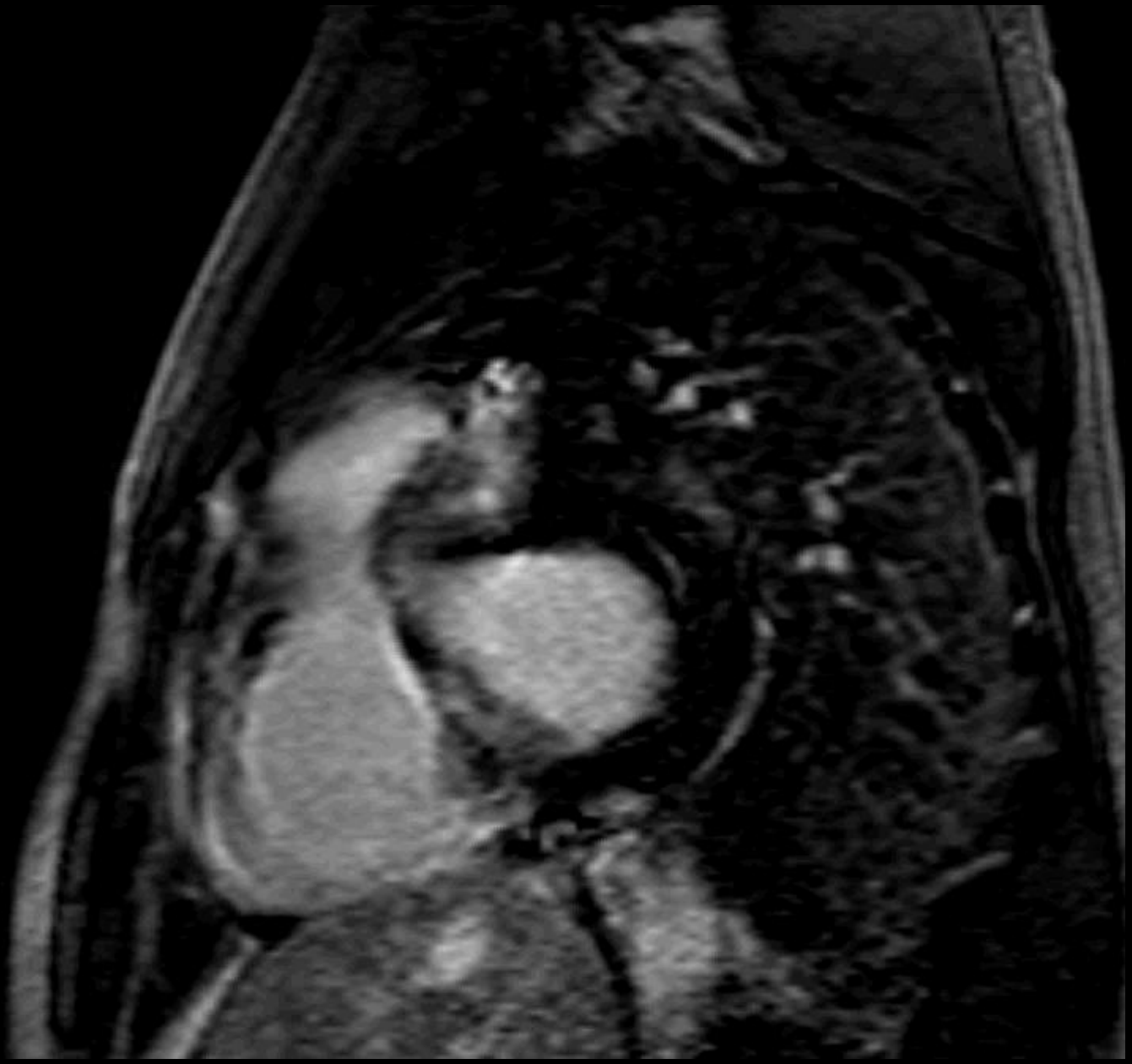
Maron et Al. JACC 2009

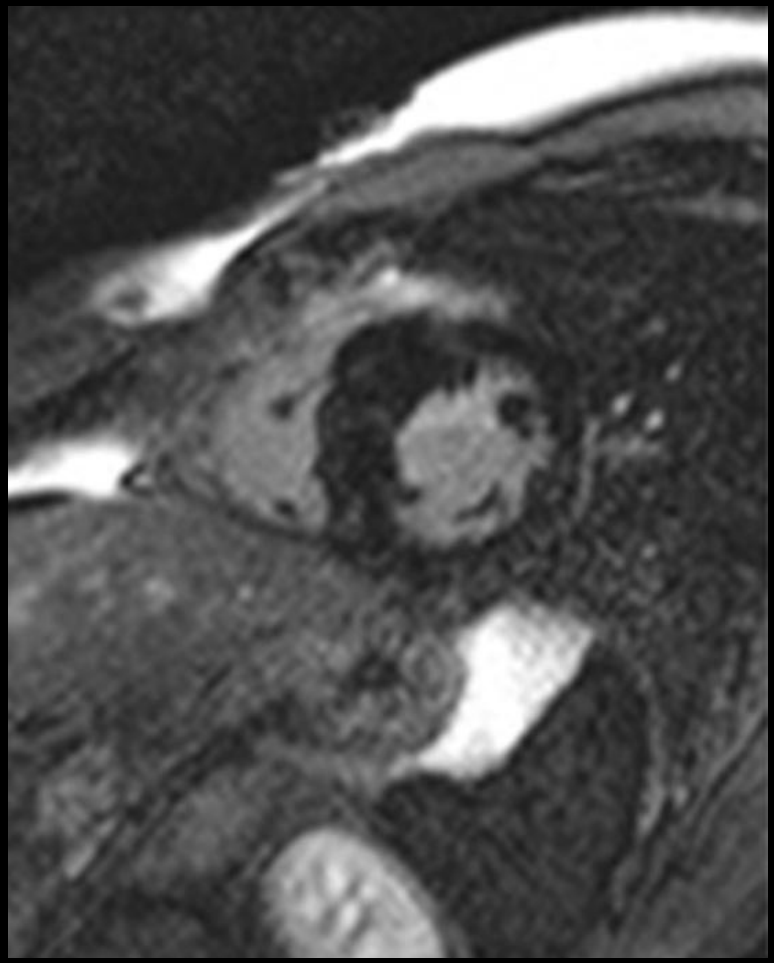
• Hypertrophic Cardiomyopathy

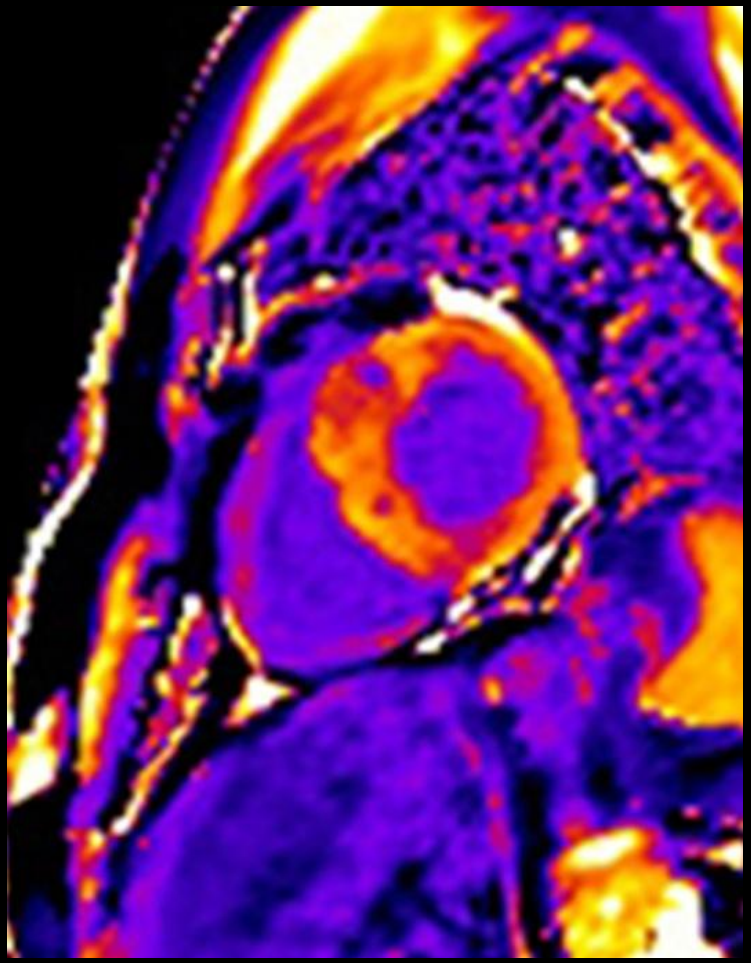


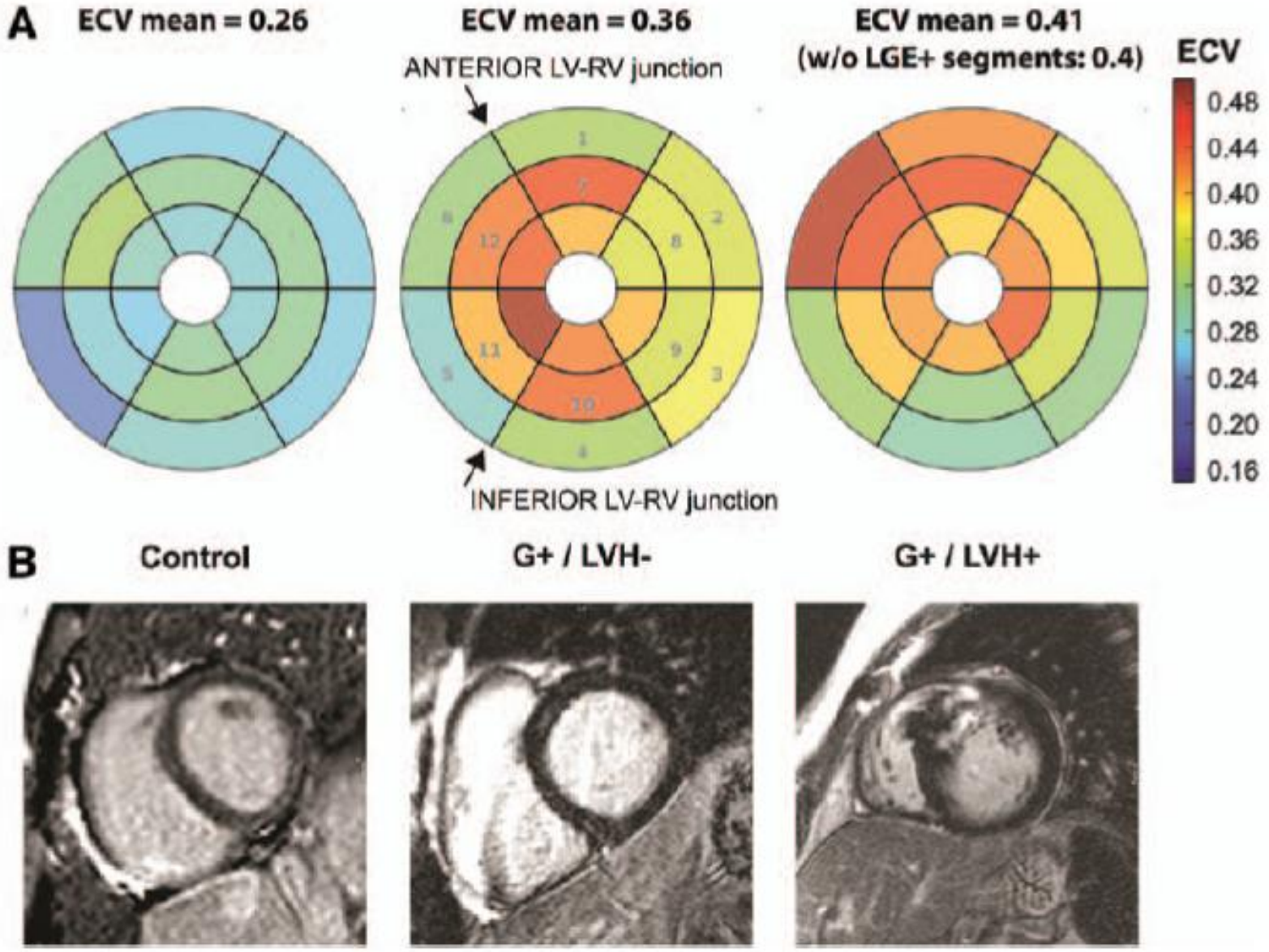












***Ho et Al. Circ Cardiovasc Imaging* 2013**

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PRACTICE GUIDELINE

2011 ACCF/AHA Guideline for the Diagnosis and Treatment of Hypertrophic Cardiomyopathy

A Report of the American College of Cardiology Foundation/
American Heart Association Task Force on Practice Guidelines

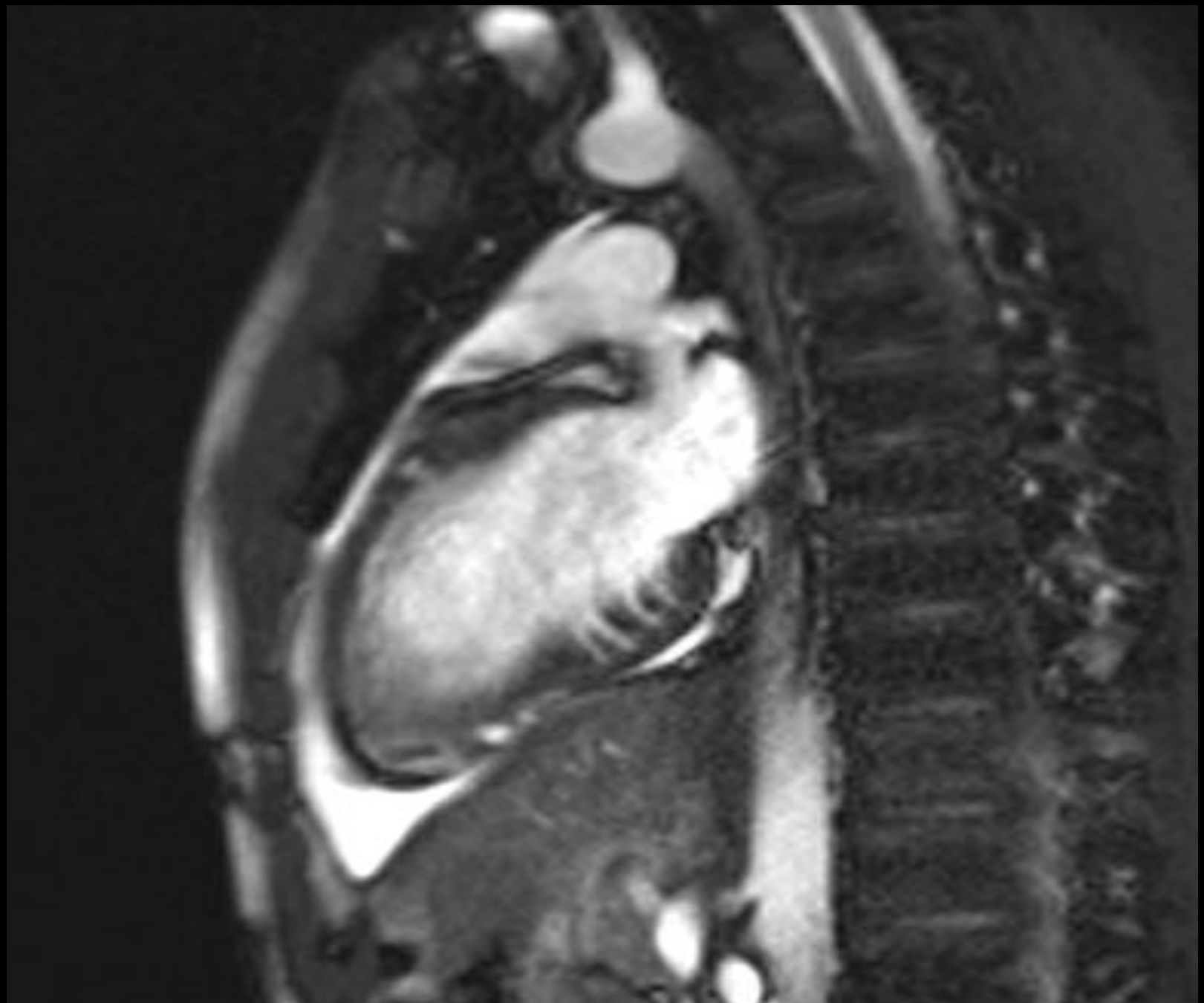
Developed in Collaboration With the American Association for Thoracic Surgery, American Society of Echocardiography, American Society of Nuclear Cardiology, Heart Failure Society of America, Heart Rhythm Society, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons

CLASS I

1. **CMR imaging is indicated in patients with suspected HCM when echocardiography is inconclusive for diagnosis (180,181). *(Level of Evidence: B)***
2. **CMR imaging is indicated in patients with known HCM when additional information that may have an impact on management or decision making regarding invasive management, such as magnitude and distribution of hypertrophy or anatomy of the mitral valve apparatus or papillary muscles, is not adequately defined with echocardiography (15,180–183). *(Level of Evidence: B)***

CLASS IIa

1. **CMR imaging is reasonable in patients with HCM to define apical hypertrophy and/or aneurysm if echocardiography is inconclusive (180,182). *(Level of Evidence: B)***



European Heart Journal – Cardiovascular Imaging
doi:10.1093/ehjci/jes005

Multiple myocardial crypts on modified long-axis view are a specific finding in pre-hypertrophic HCM mutation carriers

Wessel P. Brouwer[1,2*], Tjeerd Germans[1], Maaike C. Head[1], Jolanda van der Velden[3], Martijn W. Heymans[4], Imke Christiaans[5], Arjan C. Houweling[6], Arthur A. Wilde[7], and Albert C. van Rossum[1]

[1]Department of Cardiology, VU University Medical Center, De Boelelaan 1117, 1081 HV Amsterdam, The Netherlands; [2]Interuniversity Cardiology, Institute of the Netherlands, Utrecht, The Netherlands; [3]Department of Physiology, VU University Medical Center, Amsterdam, The Netherlands; [4]Department of Epidemiology and Biostatistics, VU University Medical Center, Amsterdam, The Netherlands; [5]Department of Clinical Genetics, Academic Medical Center, Amsterdam, The Netherlands; [6]Department of Clinical Genetics, VU University Medical Center, Amsterdam, The Netherlands; and [7]Department of Cardiology, Academic Medical Center, Amsterdam, The Netherlands

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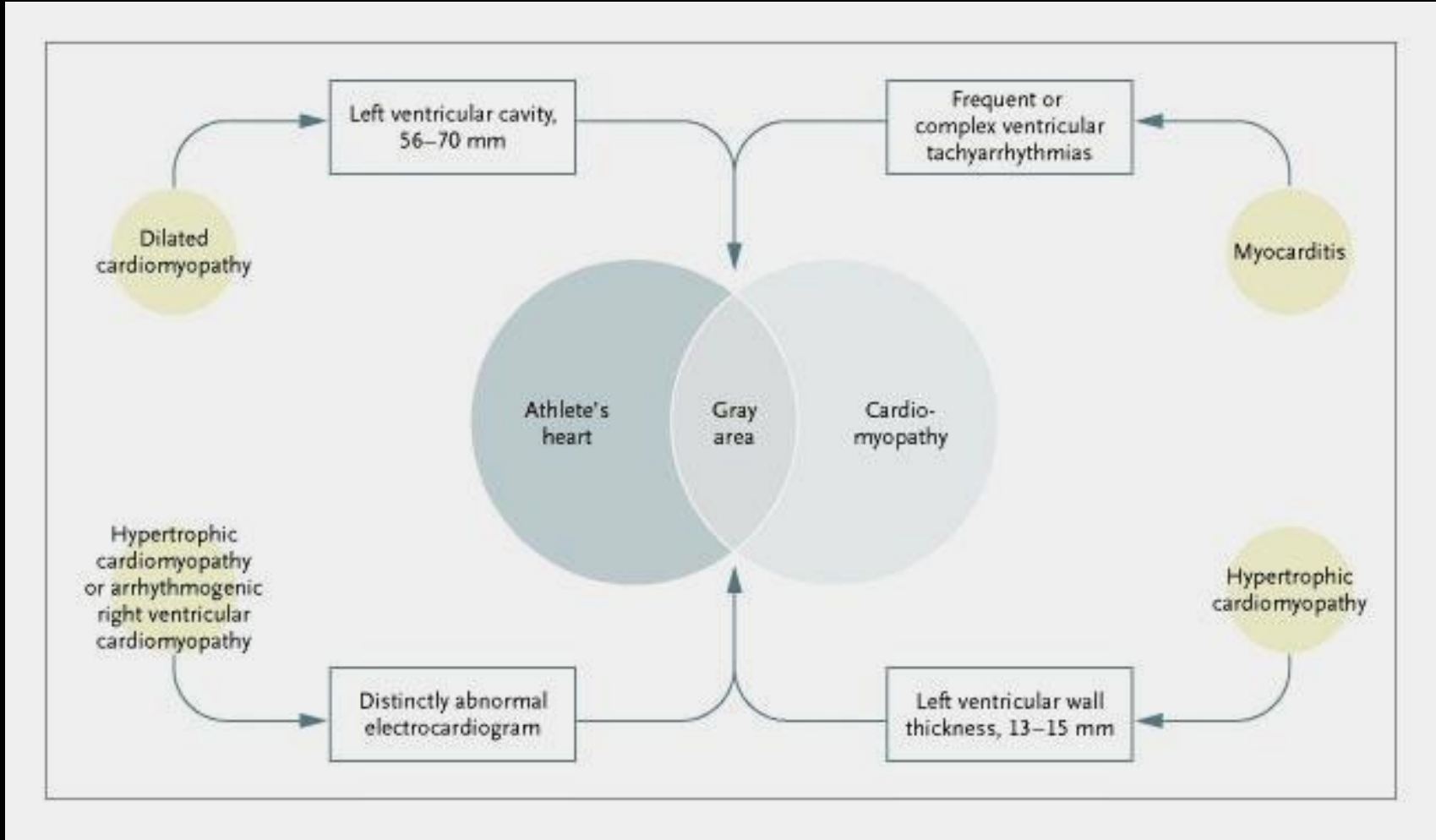
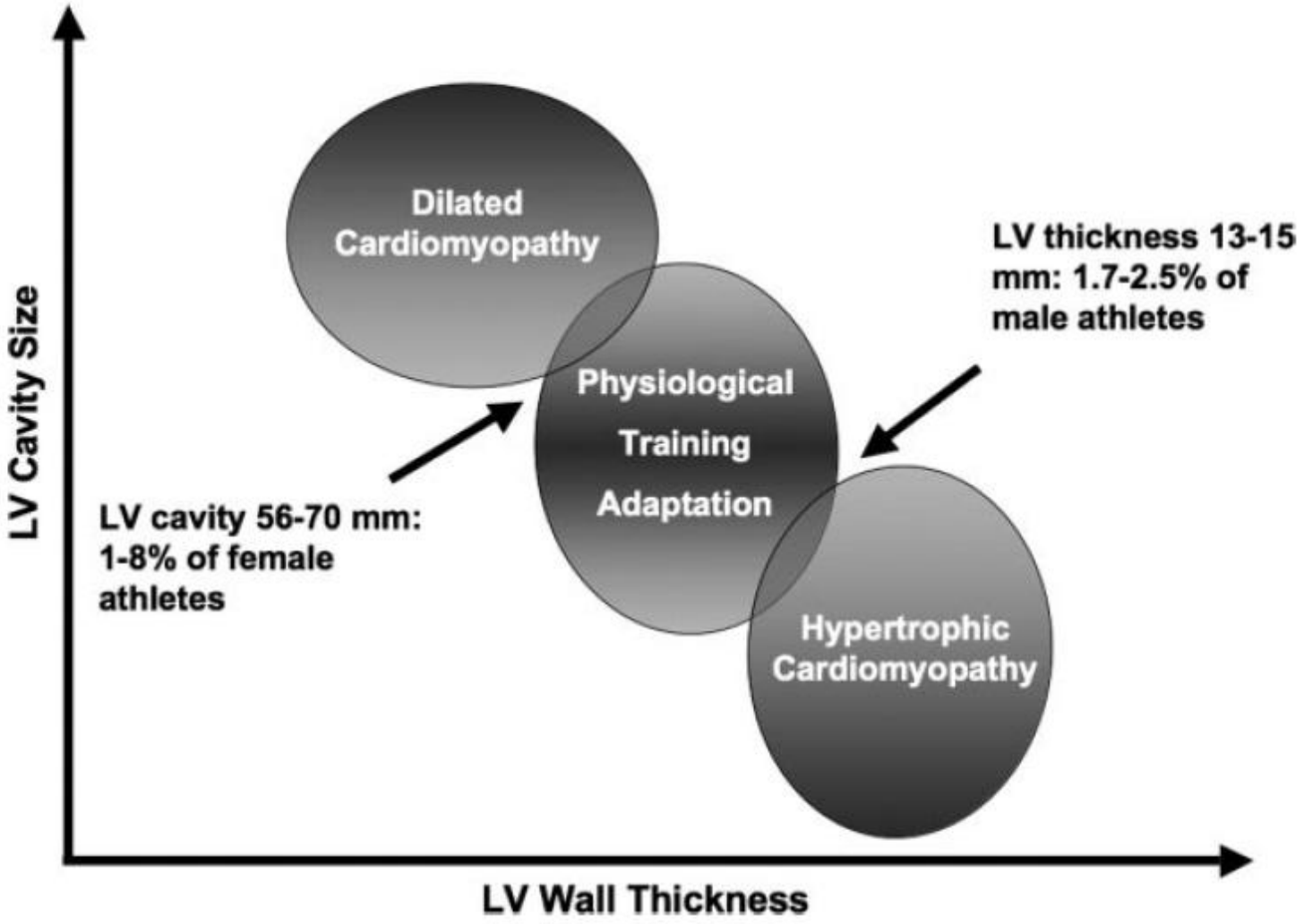
Zona grigia tra Cuore d'Atleta e Cardiomiopatia Ipertrofica



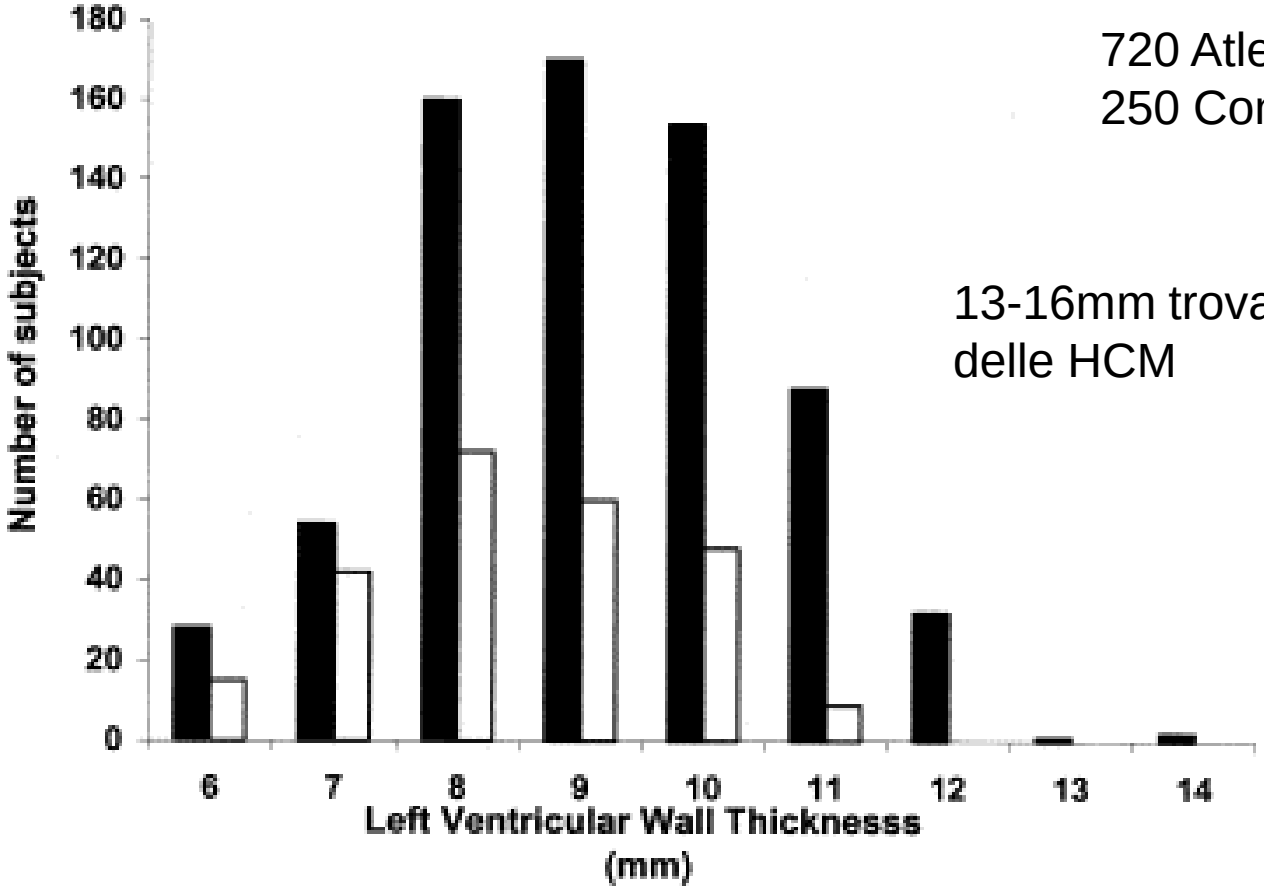
Maron B. N Engl J Med 2003

Does Size Matter? Clinical Applications of Scaling Cardiac Size and Function for Body Size



Dewey et Al. Circulation 2008

Ipertrofia dell'Atleta e HCM

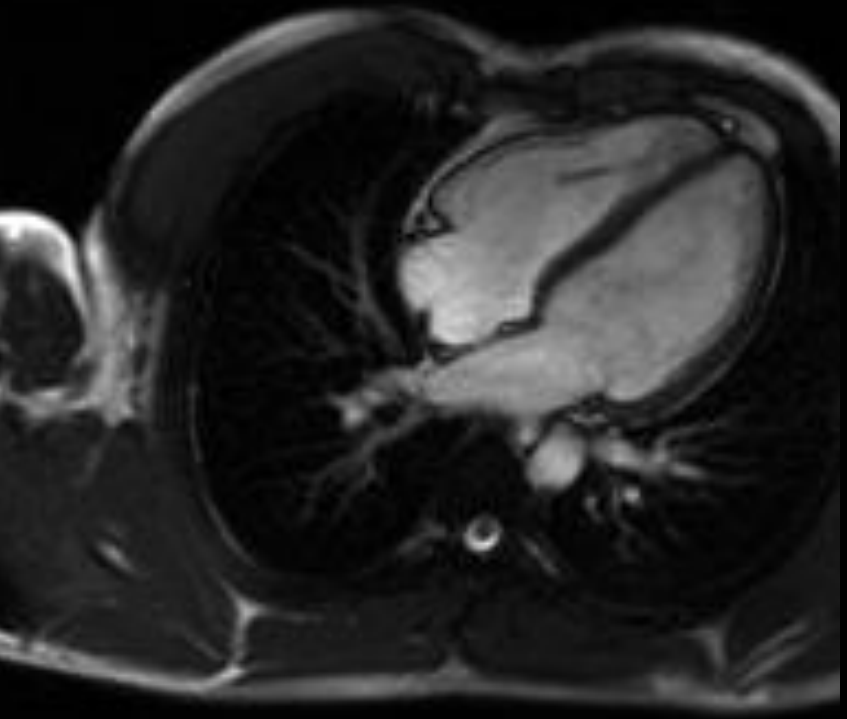


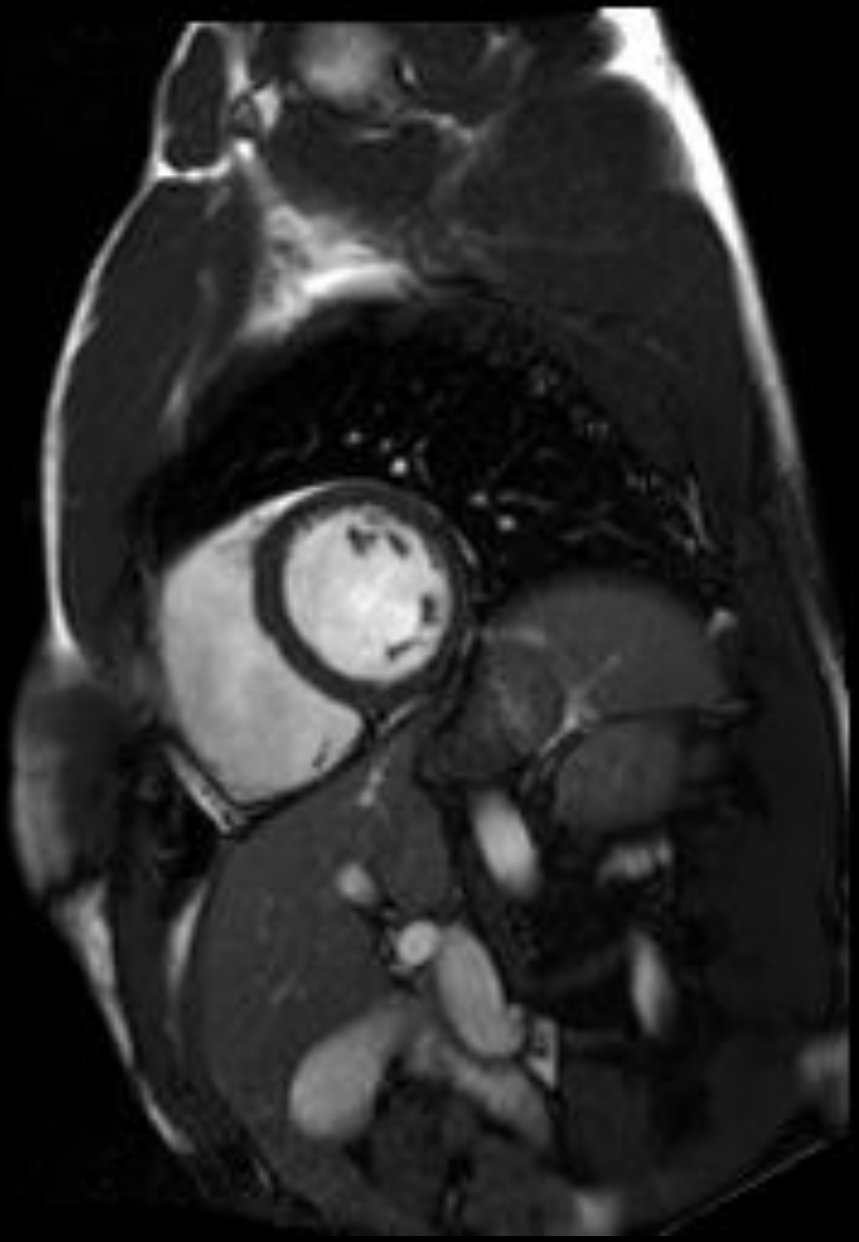
720 Atleti
250 Controlli

13-16mm trovati nel 10-15% delle HCM

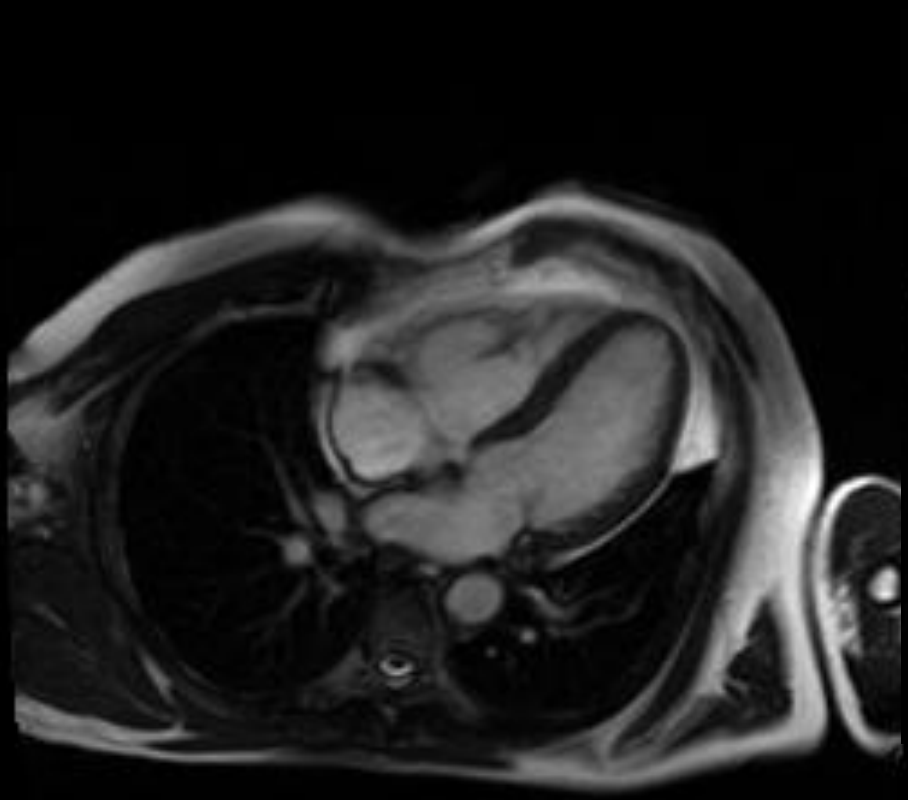
Sharma et Al. JACC 2002

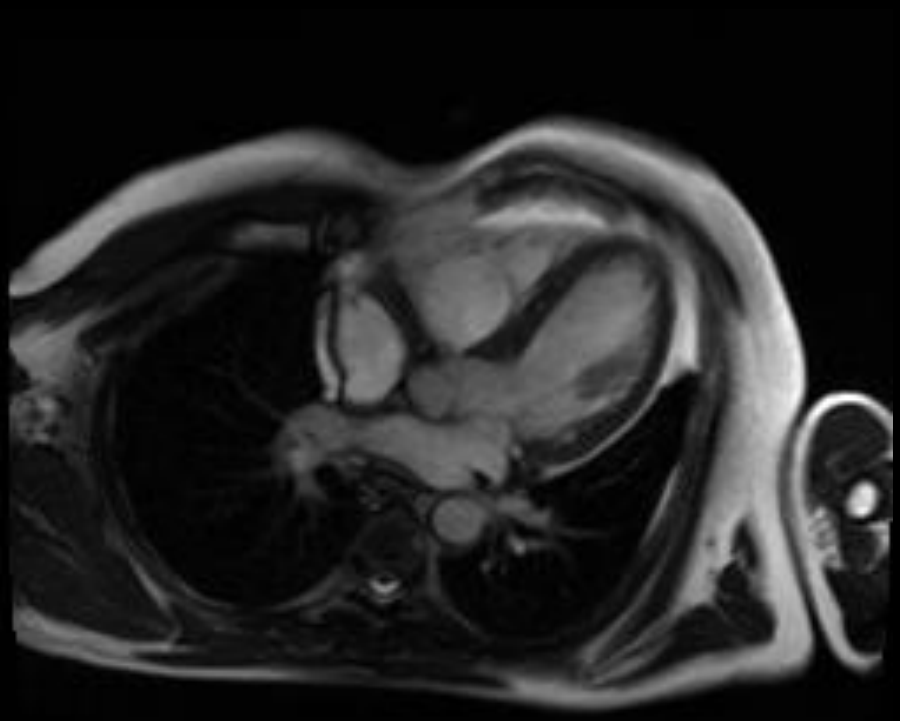
Hockey su prato



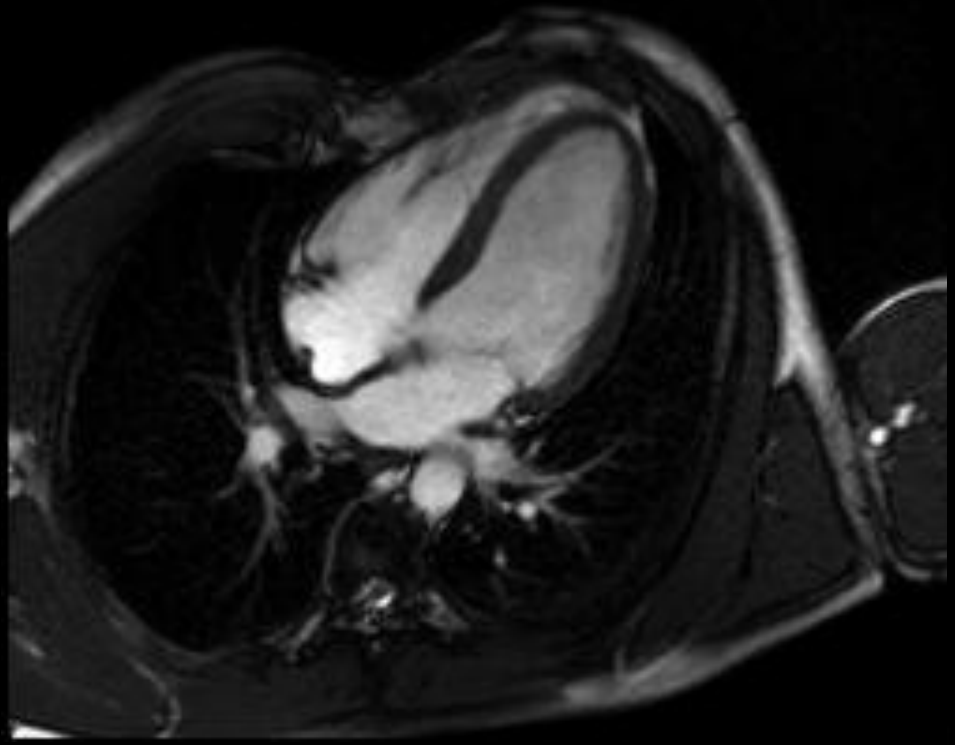


Nuoto agonistico



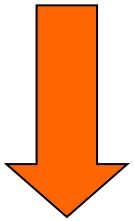


Ciclismo agonistico



RM e ruolo nella CMP Ipertrofica

- Accurata definizione del miocardio
- Precisa misurazione degli spessori e della massa (modificazioni sisto-diastoliche)



Gold standard per le altre metodiche

Original Article

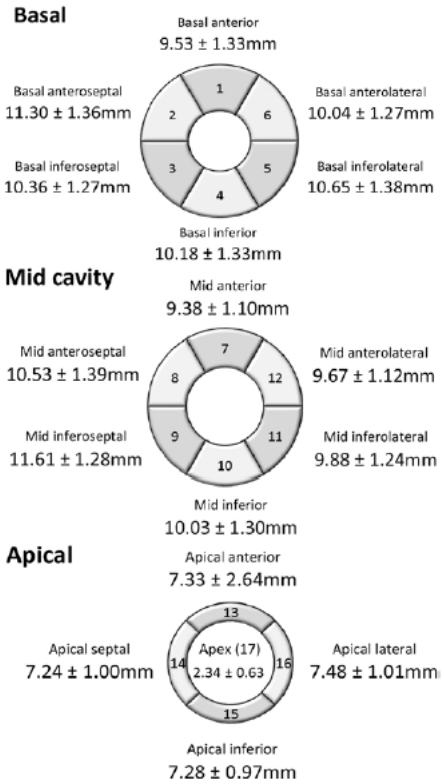
Left Ventricular Wall Thickness and the Presence of Asymmetric Hypertrophy in Healthy Young Army Recruits

Data From the LARGE Heart Study

Phong T. Lee, MD*; Marc R. Dweck, MD*; Sparsh Prasher, MD; Anoop Shah, MD; Steve E. Humphries, MD; Dudley J. Pennell, MD; Hugh E. Montgomery, MD; John R. Payne, MD

541 Reclute
CMR

23% had a maximal wall thickness ≥13.0 mm, whereas the prevalence of asymmetrical wall thickening increased from 2.2% to 10% after an exercise-training program



Lee et Al. ***Circ Cardiovasc Imaging.*** **2013;6:262-267**

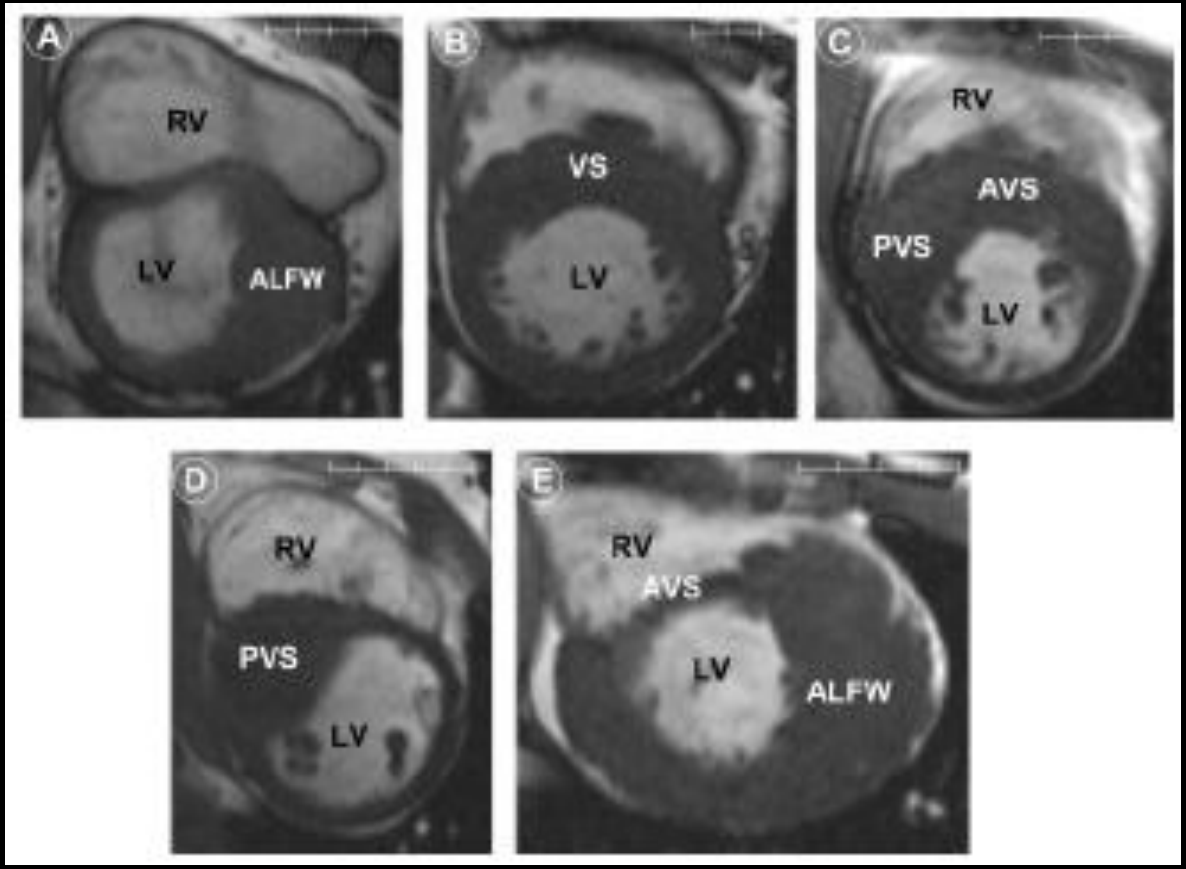
A chi riservare la RM?

- A tutti gli Atleti?

- Ad Atleti con spessore >12mm e/o con ecocardiogramma dubbio?

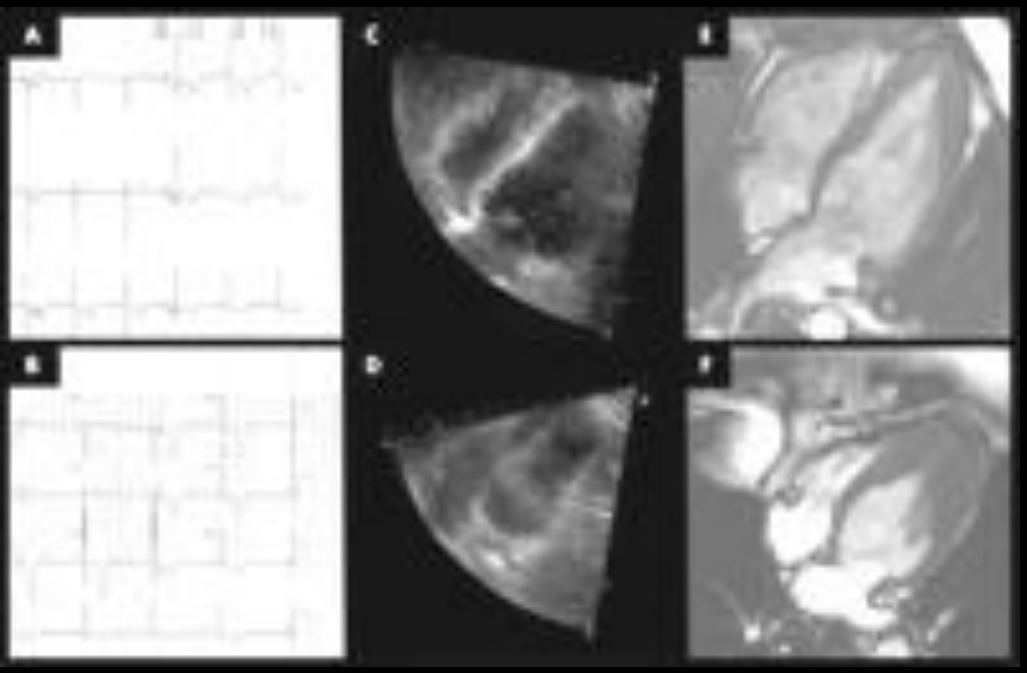
- Ad Atleti con alterazioni ECG anche in assenza di alterazioni all'eco?

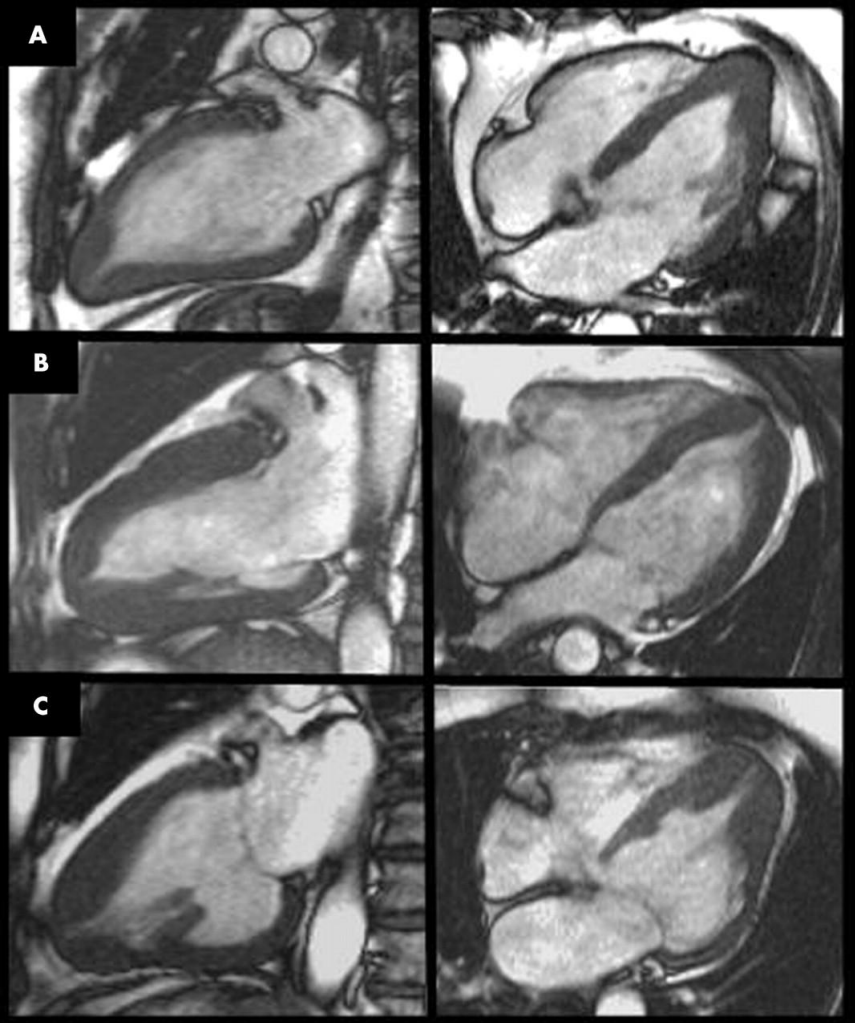
Diagnosi di HCM non rilevata all'ecocardiografia



Rickers et Al. *Circulation*. 2005;112:855-861

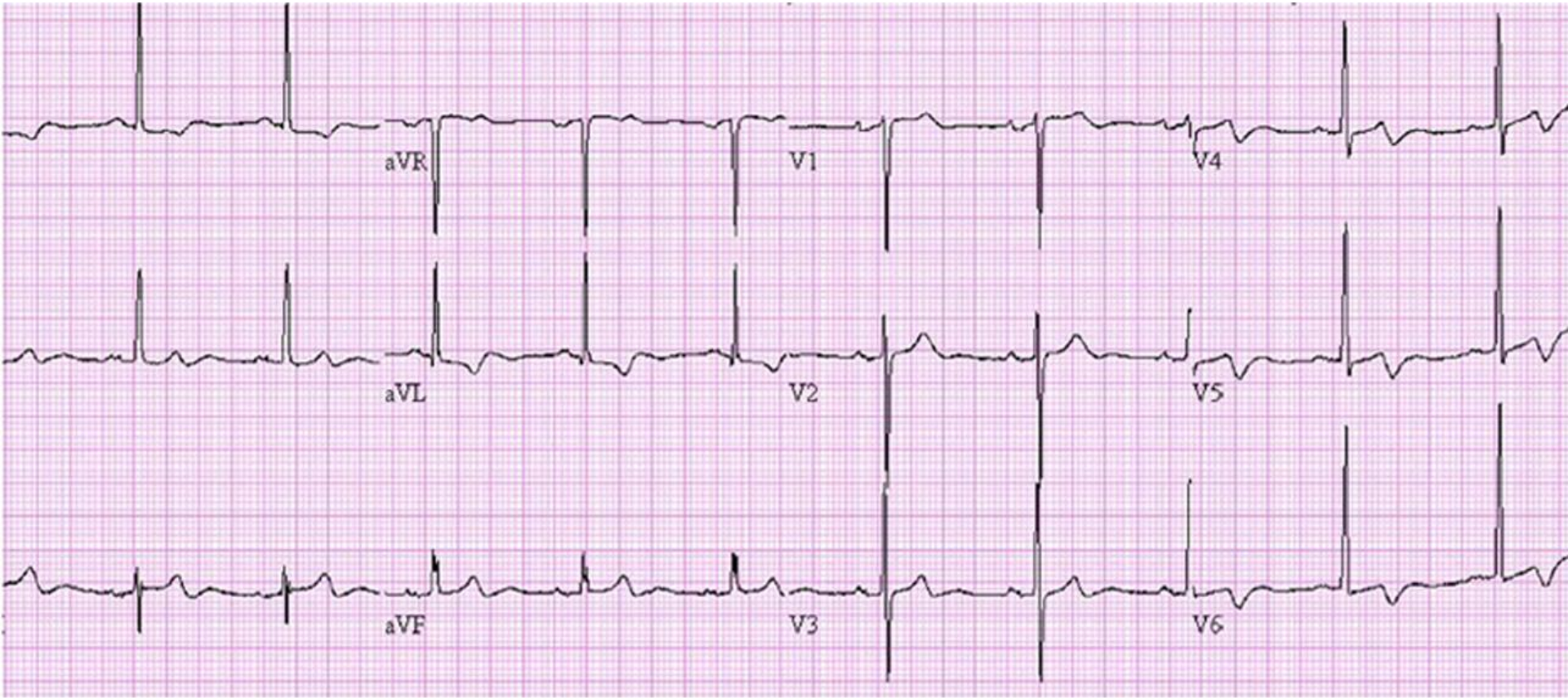
Diagnosi di HCM non rilevata all'ecocardiografia

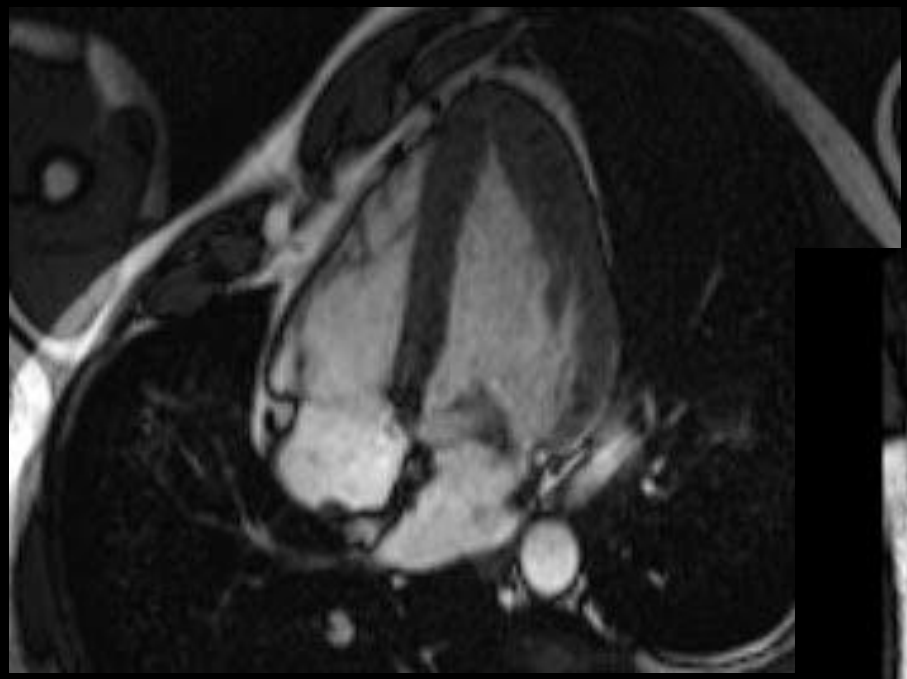


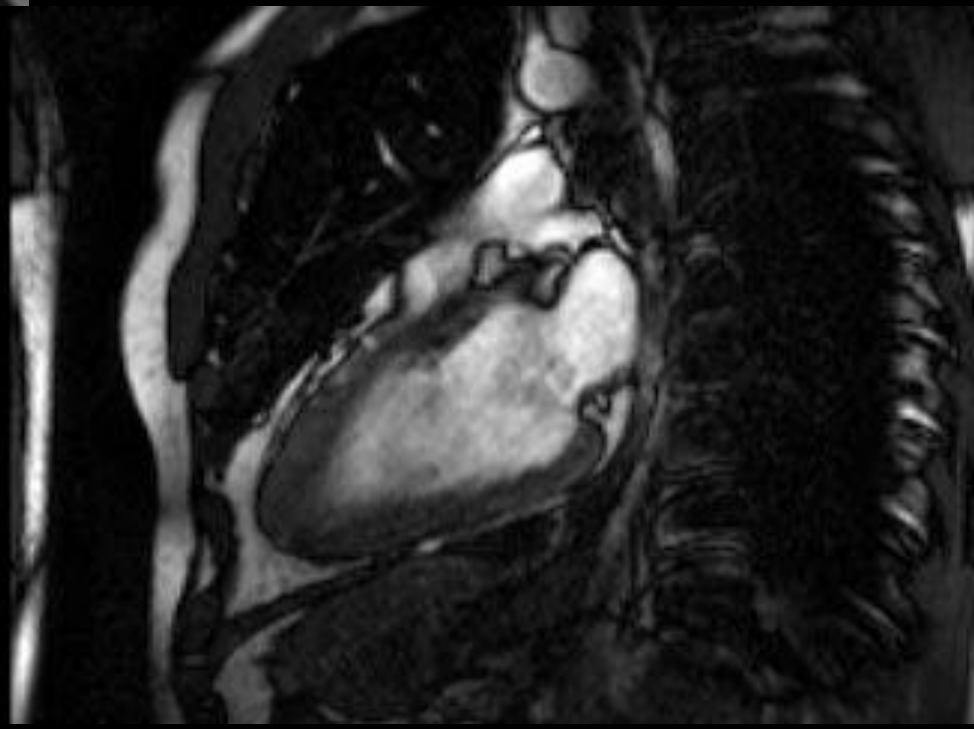


Moon et Al. Heart 2004; **90**:645-649

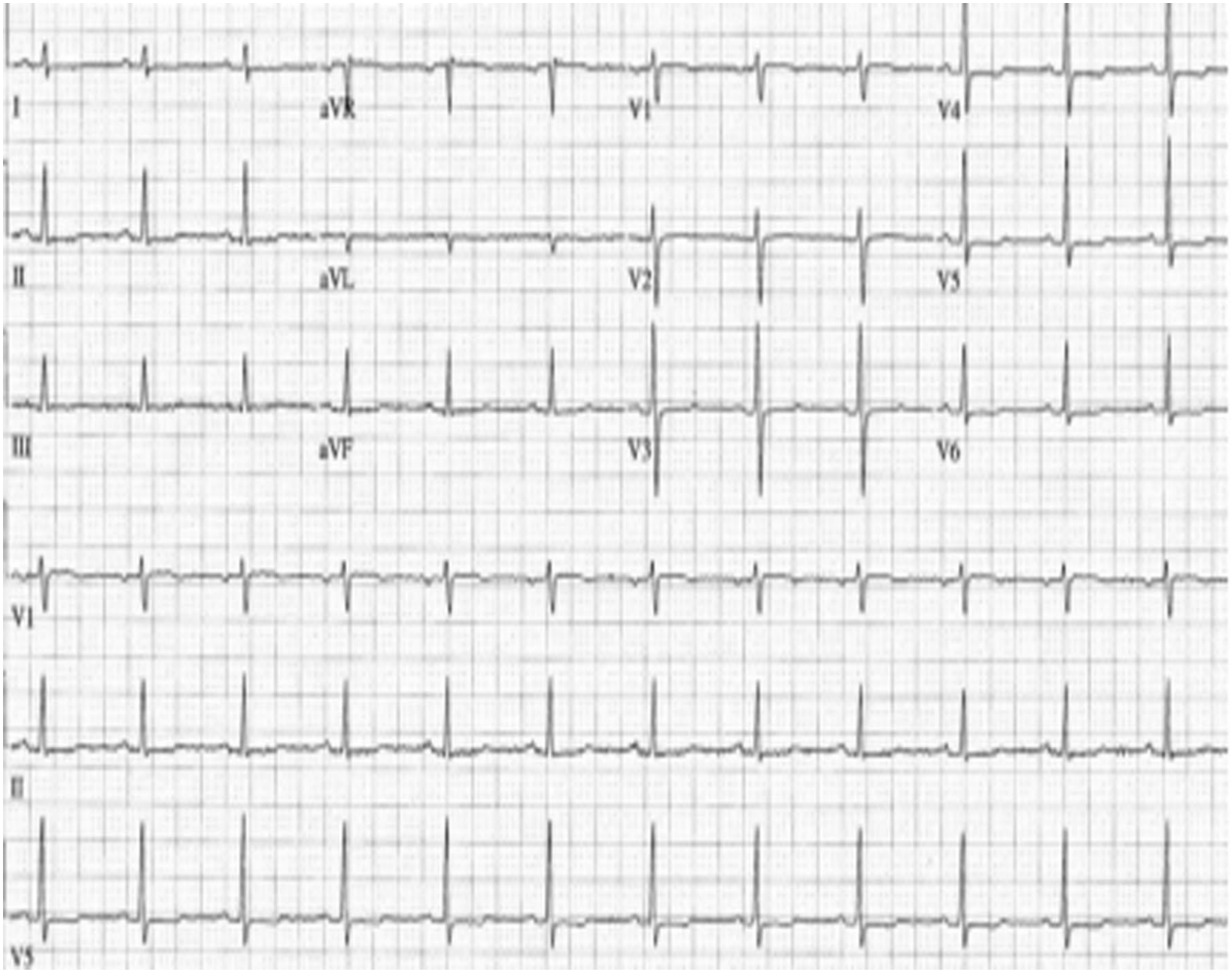
Atleta con alterazioni ST-T

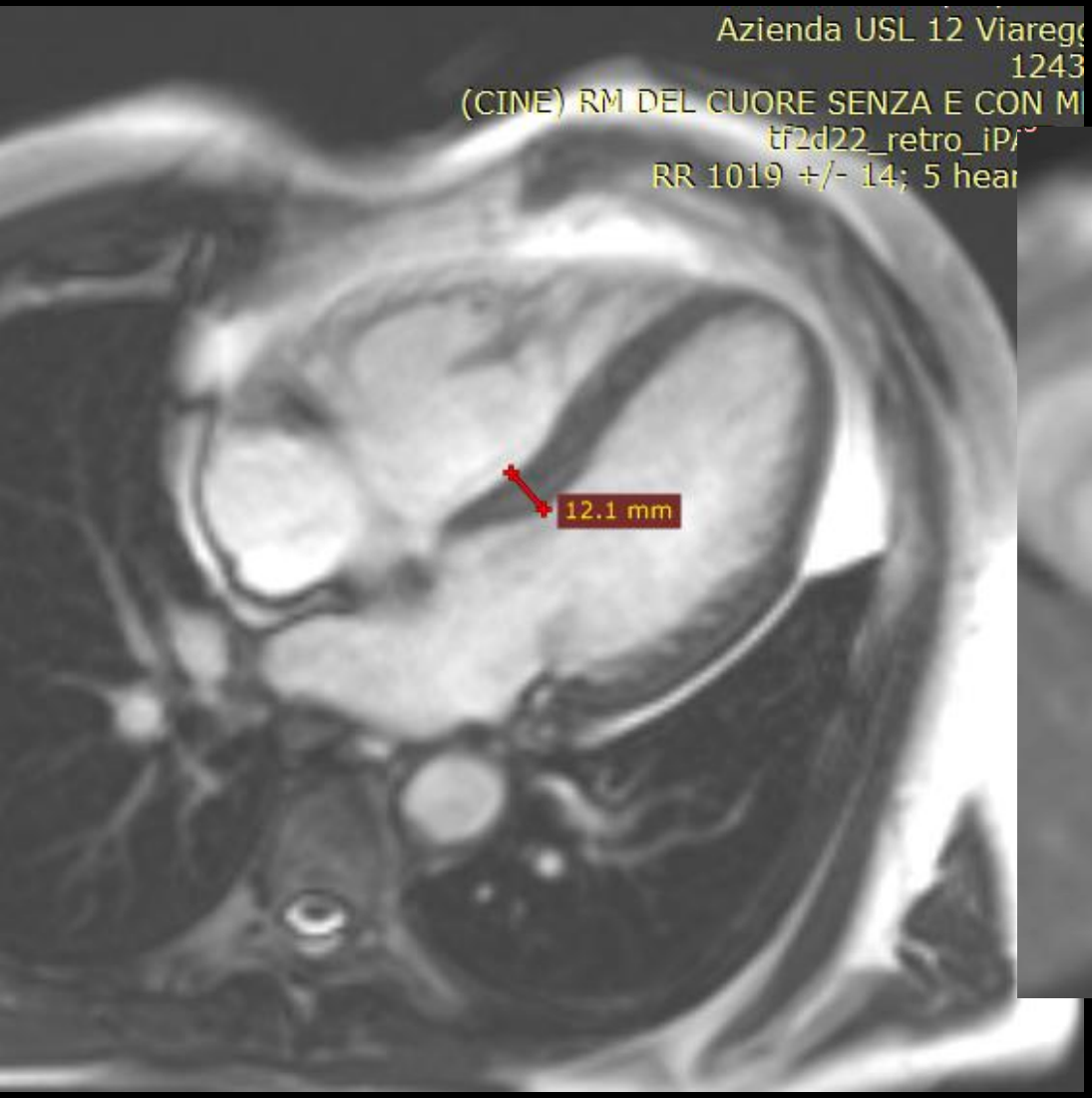






LGE in paziente con HCM apicale


I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
V1
II
V5


Azienda USL 12 Viareg
1243
(CINE) RM DEL CUORE SENZA E CON M
tf2d22_retro_iP
RR 1019 +/- 14; 5 hea
12.1 mm


SA
(CINE)
12.3 mm

Quando ricorrere alla RM ?

- In tutti i casi in cui vi sono alterazioni ECG che non sono comuni negli atleti
- Negli atleti che mostrano una ipertrofia di parete incongrua rispetto all'attività sportiva praticata
- In coloro che praticano attività sportiva ed hanno familiarità per CMP
- In presenza di atleti con ipertrofia e presenza di aritmie ventricolari

Aspetti suggestivi per CMP ipertrofica

Symptoms	Unexplained syncope—particularly during exercise
	Palpitations
	Shortness of breath disproportionate to the exercise performed
	Dizziness
	Chest pain
Family history	HCM in a first-degree relative
Demographics	Age < 16 years old
	Female sex
	Participation in purely isometric sport
	Small body surface area
Echocardiography	Left ventricular wall thickness > 16 mm
	Asymmetrical septal hypertrophy
	Small left ventricular cavity diameter in end-diastole
	Presence of systolic anterior motion of the mitral valve leaflet and associated left ventricular outflow obstruction
	Abnormal indices of diastolic function
12-Lead ECG	Pathological Q-waves
	ST segment depression
	Left bundle branch block
	T-wave inversions in the lateral/inferior leads
Cardiopulmonary exercise testing	Peak $V_{O2\ max}$ < 50mL/kg/min or < 120% of predicted maximum
Cardiac MRI	Demonstration of apical hypertrophy
	Demonstration of significant myocardial fibrosis with gadolinium enhancement
Detraining	Failure of regression of left ventricular hypertrophy

Rawlins et Al. European Journal of Echocardiography (2009)

Conclusioni

- La diagnosi di CMP ipertrofica poggia su elementi non solo morfologici di cui occorre tener conto nel momento in cui si ritiene che la RM sia utile
- Esiste tuttora incertezza su quali siano i limiti fisiologici dell'ipertrofia dell'Atleta
- L'attività fisica strenua può svelare una cardiomiopatia sottostante
- La frequenza della HCM è di 1:500 nella popolazione generale
- La RM è una tecnica molti importante nella diagnostica delle CMP ed in particolare della HCOM

Highlights

- Esiste una ipertrofia normale?
- Come si definisce l'ipertrofia dell'atleta?
- Quando si parla di cardiomiopatia ipertrofica?
- Esiste una sola cardiomiopatia ipertrofica?
- Quali tecniche si impiegano per valutare la ipertrofia (dell'atleta, ipertrofica, altro?)

Valori “normali” di massa miocardica

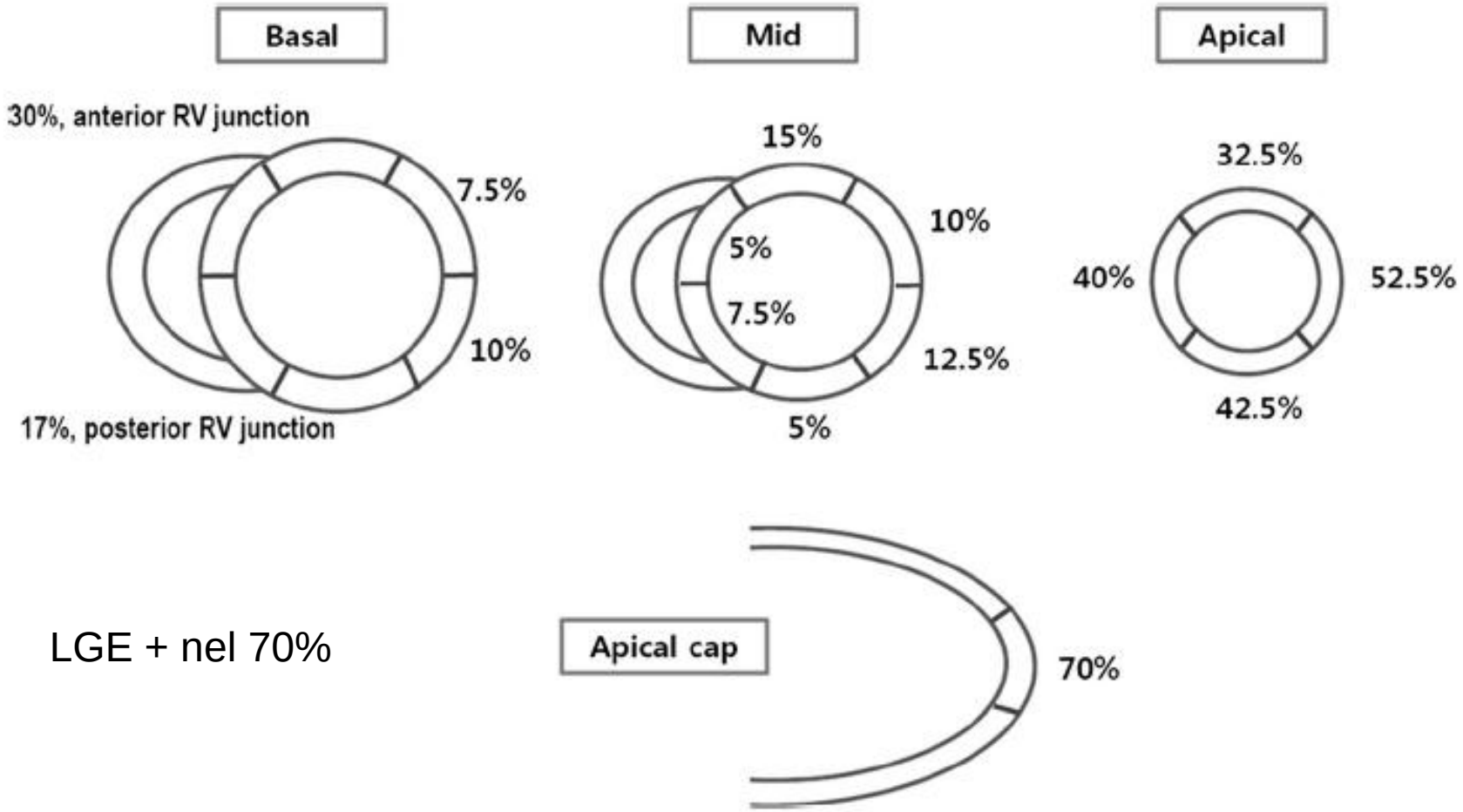
- Ampiamente variabili
- Dipendenza da più fattori (anche dinamici)
  - Età
  - Sesso
  - Superficie corporea

- Rapporto non diretto con gli spessori
- Dipendenza dal carico

Discriminare il cuore d'atleta dalla cardiomiopatia ipertrofica

- Differenziare l'ipertrofia compensatoria da quella patologica

- Identificare elementi peculiari
  - Anatomici
  - Funzionali
  - Isto-patologici


Basal
Mid
Apical
30%, anterior RV junction
7.5%
10%
17%, posterior RV junction
15%
10%
5%
7.5%
12.5%
5%
32.5%
40%
52.5%
42.5%
Apical cap
70%

LGE + nel 70%