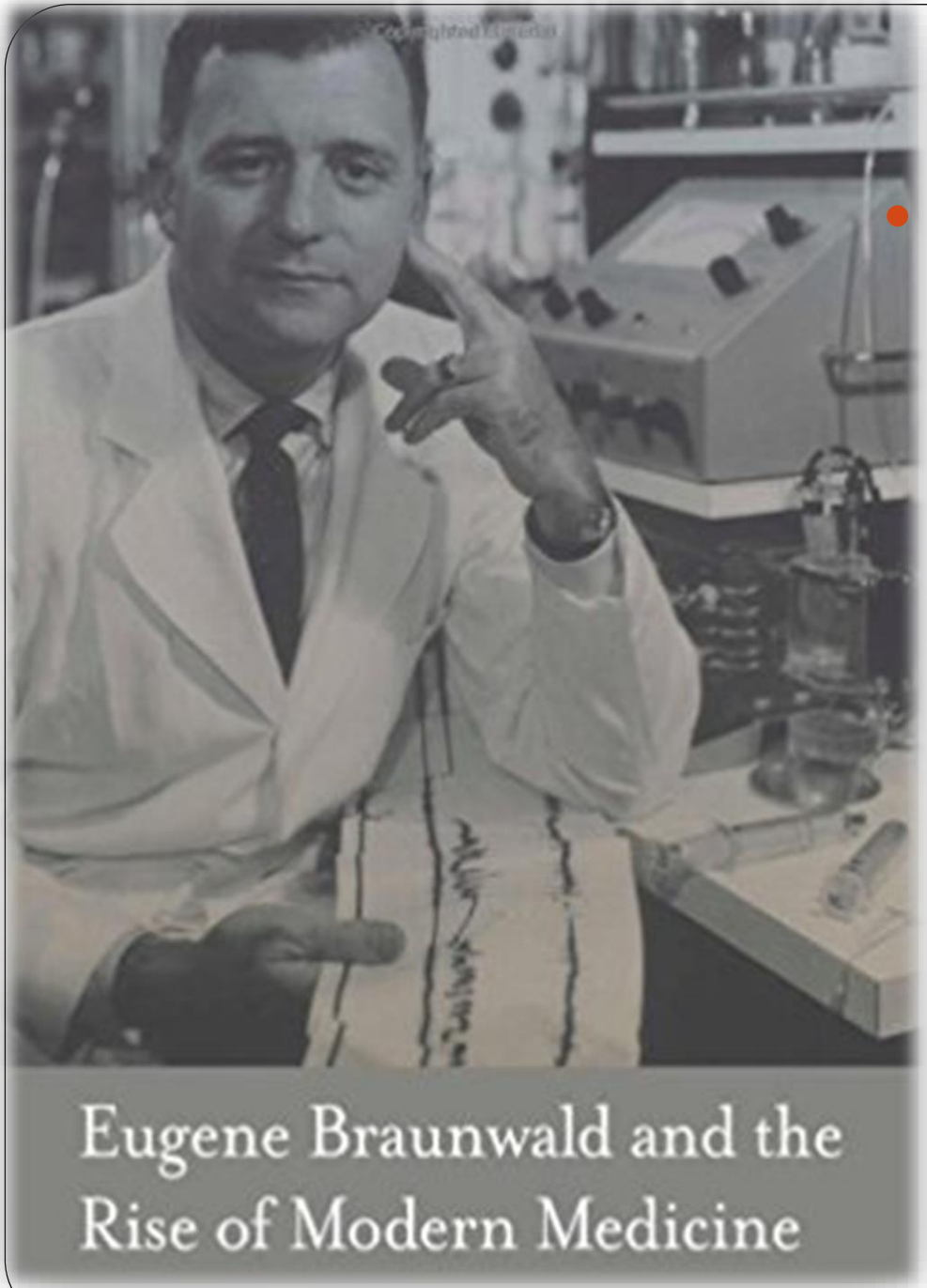
ROLE OF CARDIAC MAGNETIC RESONANCE IN DIAGNOSIS AND PROGNOSIS OF CARDIOMYOPATHY

TRẦN THỊ XUÂN ANH. MD

CARDIOLOGY DEPARMENT

UNIVERSITY MEDICAL CENTER HCM



- "Increasingly, cardiology is a visual field, so recognizing tracings, recognizing images and knowing how to use MRIs is a challenge and a skill that need to be practiced".

Dr Eugene Braunwald

CARDIAC MRI STUDY

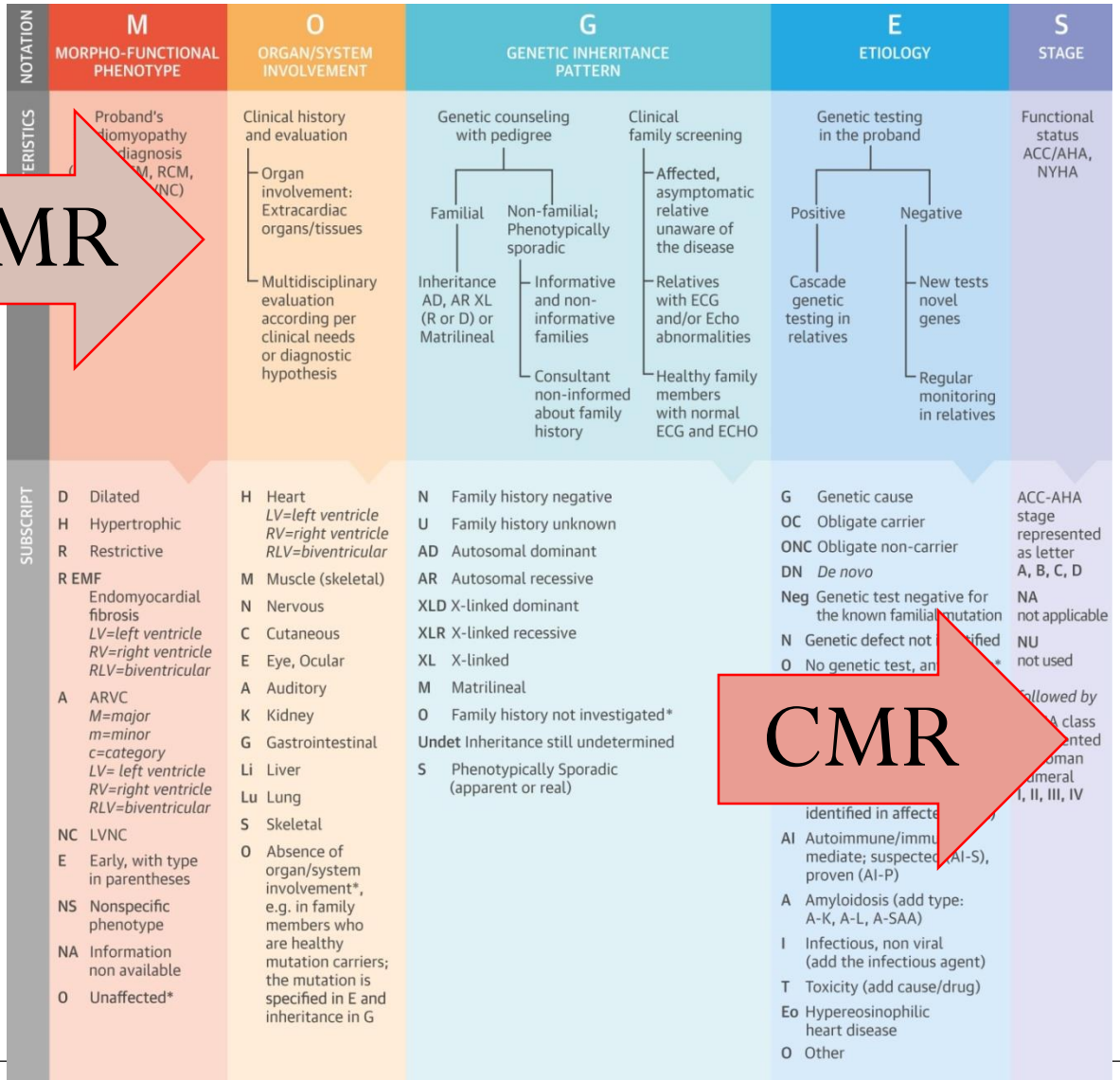
- Cadiac Anatomy -Congenital Heart Disease
- Ventricular Function
- Differentiation of ischemic and nonischemic .
- Cardiomyopathies
- Cardiac masses
- Pericardial disease
- Valvular disease
- Coronary artery disease
- Pulmonary vein assessment

DEFINICATION OF CARDIOMYOPATHIES

1995 World Health Organization/International Society and Federation of Cardiology

Cardiomyopathies are defined as diseases of the myocardium associated with cardiac dysfunction.

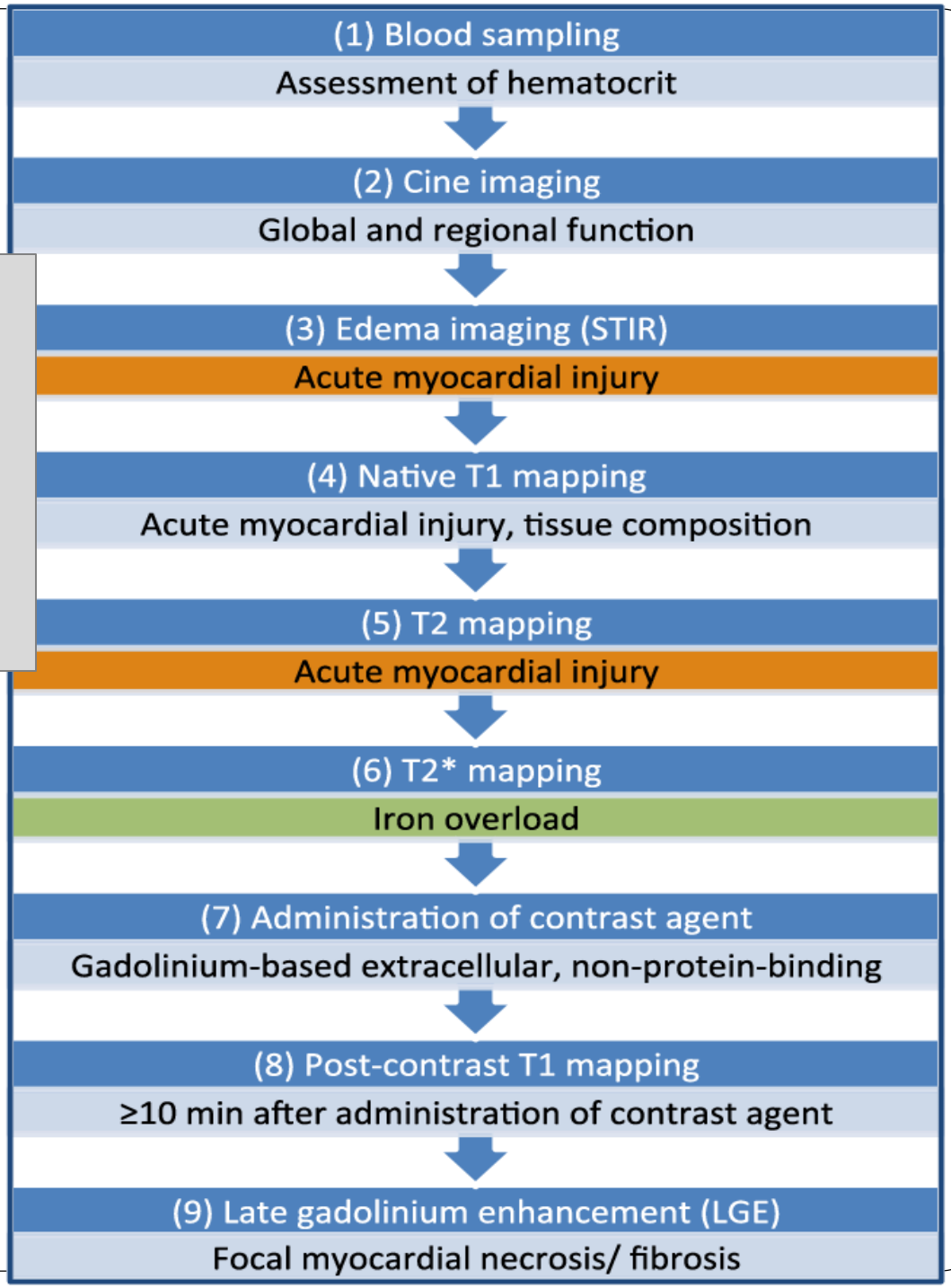
MOGE(S) CLASSIFICATION OF CARDIOMYOPATHIES-WHF 2013



CARDIAC MRI PROTOCOL FOR MYOCARDIAL CHARACTERIZATION

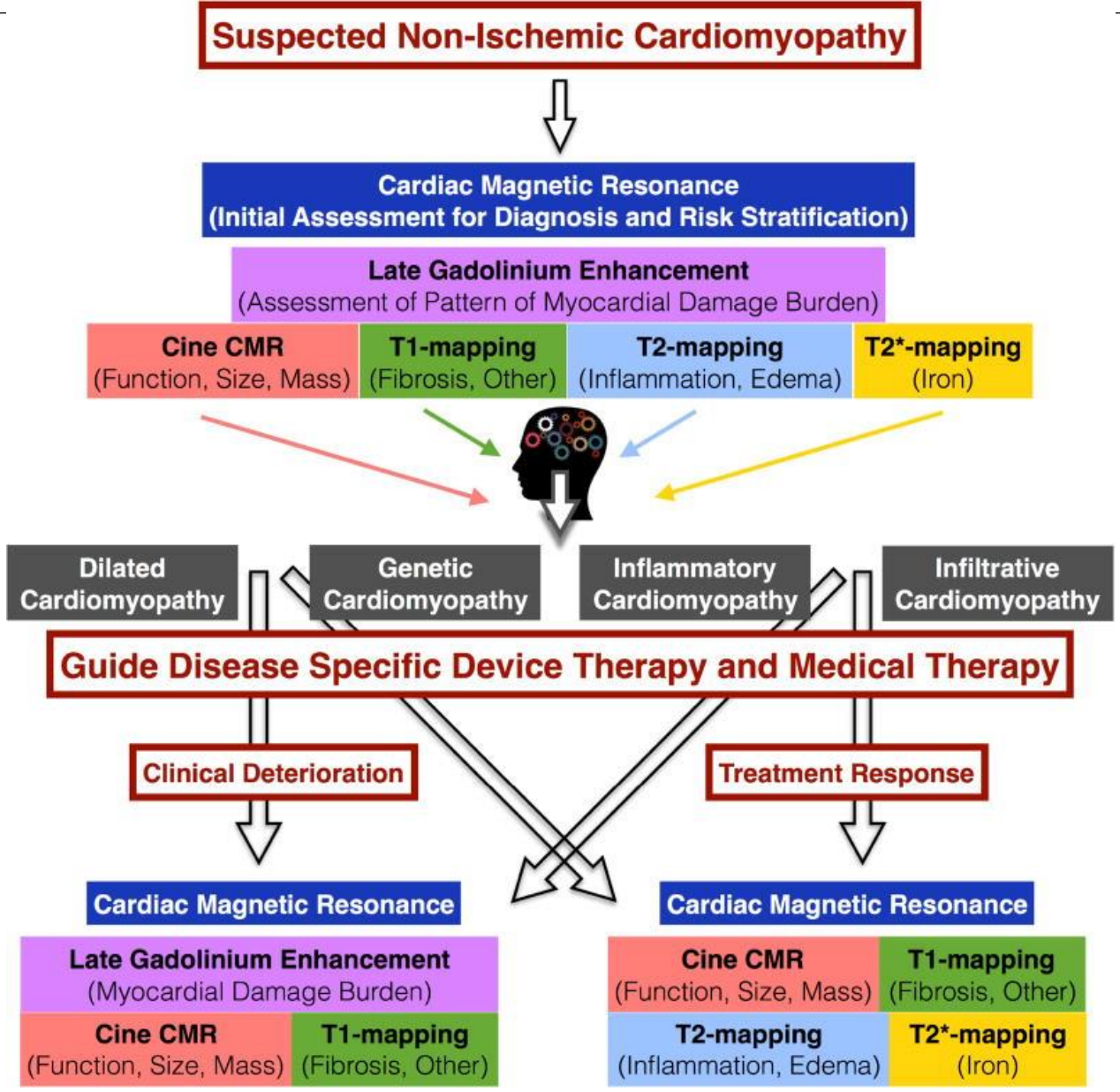
Early enhancement (first 5 min after injection Gd)

Late enhancement (5 to 20 min after injection Gd)



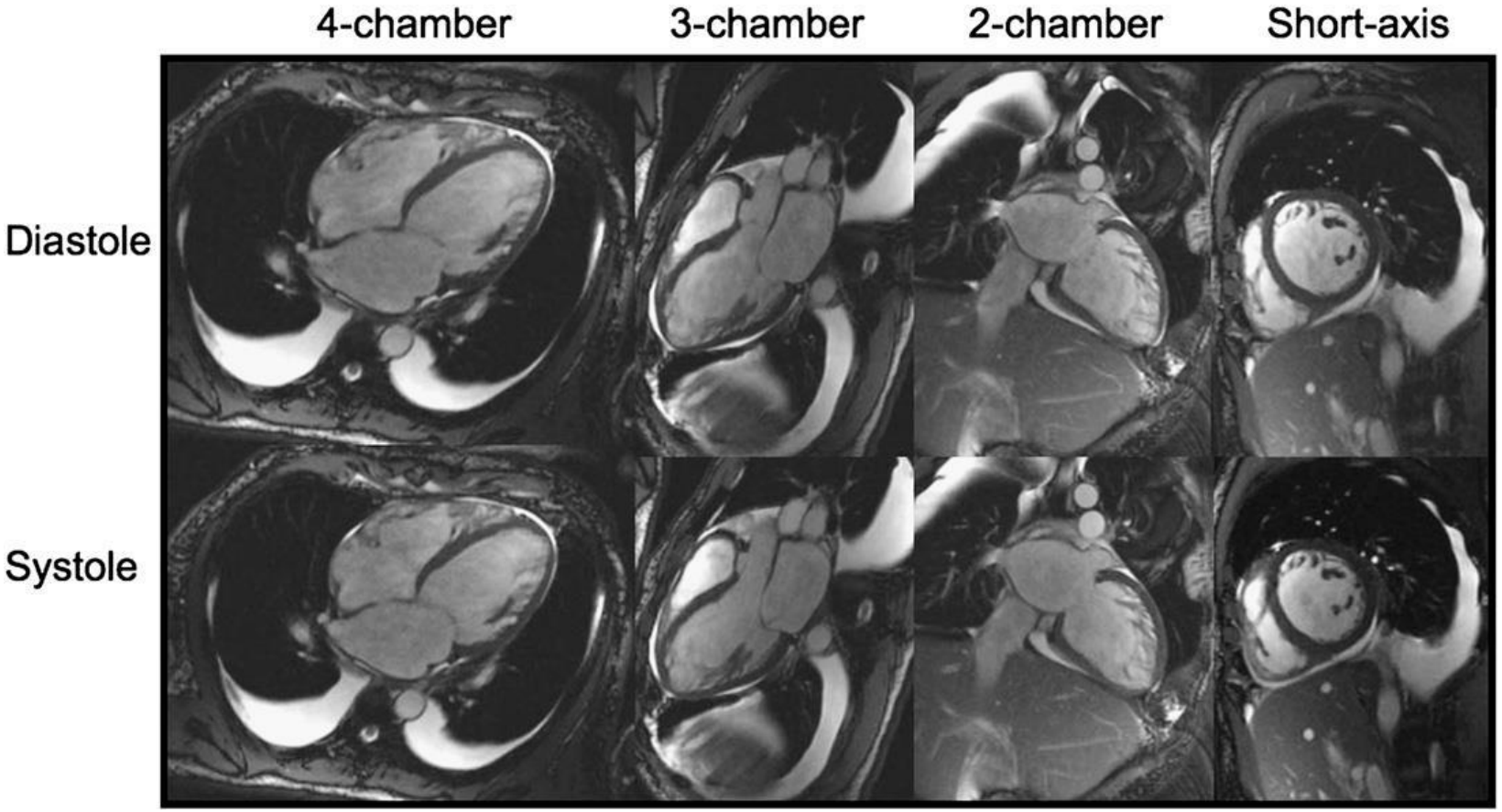
CMR Report

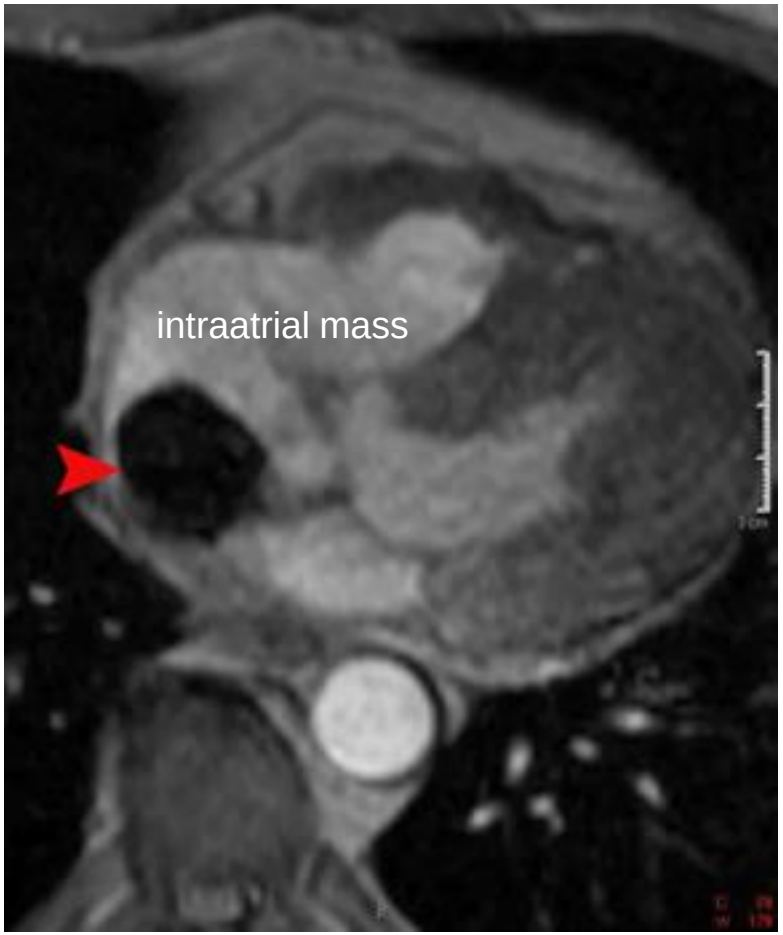
- **Dimensions, mass** (corrected for BSA) Thickness of interatrial septum, ventricle wall.
- **Function**
  - LV: EDV, ESV , SV, EF, longitudinal function, mass
  - RV: EDV, ESV, SV, EF, longitudinal function
  - Regional wall motion abnormalities
- **Valve regurgitation**
- **LGE pattern**
- **Pericardial / pleural effusion.**
- **Cardiac ion overload.**



CENTRAL ILLUSTRATIONEvaluation of Nonischemic Cardiomyopathy Using CMRPatel, A.R. et al. J Am Coll Cardiol Img. 2017;10(10):1180–93

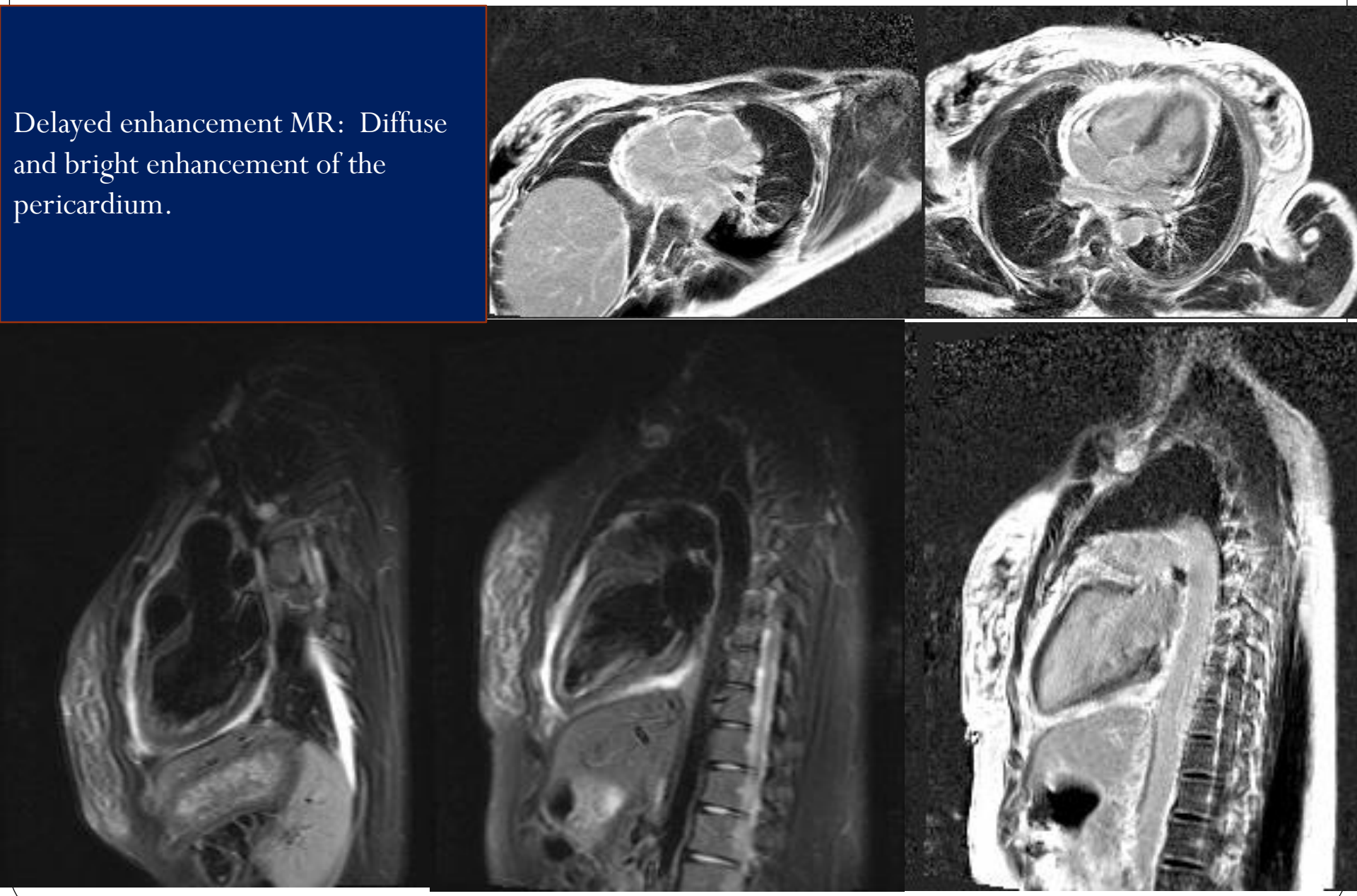
Cine image

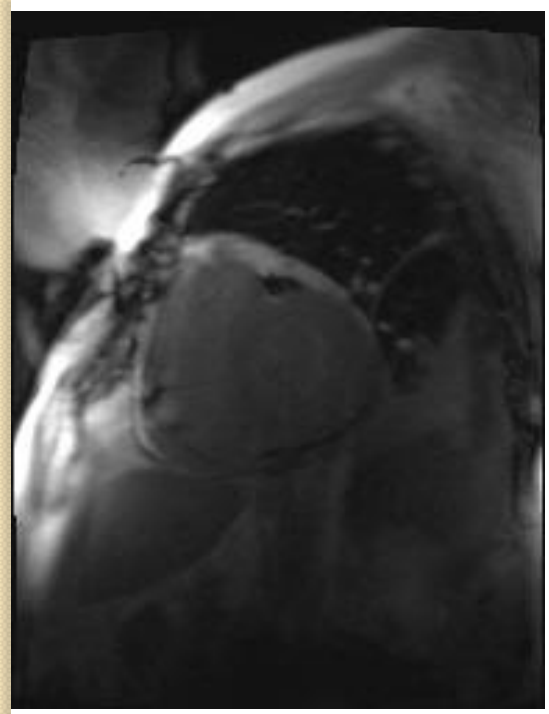


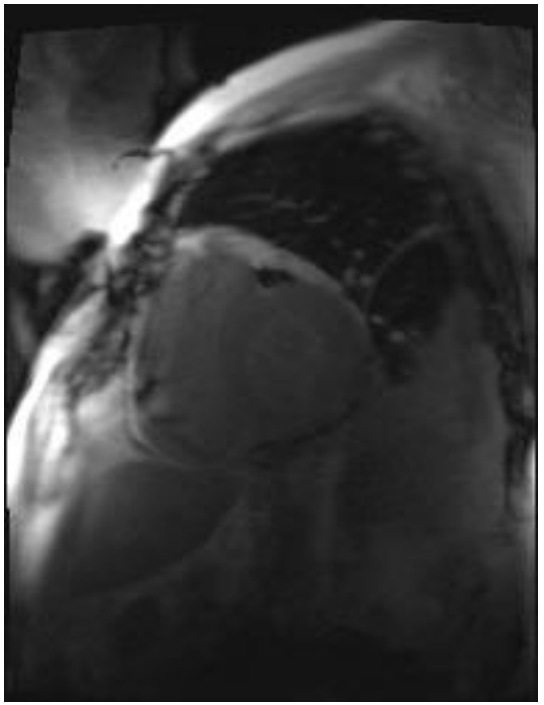


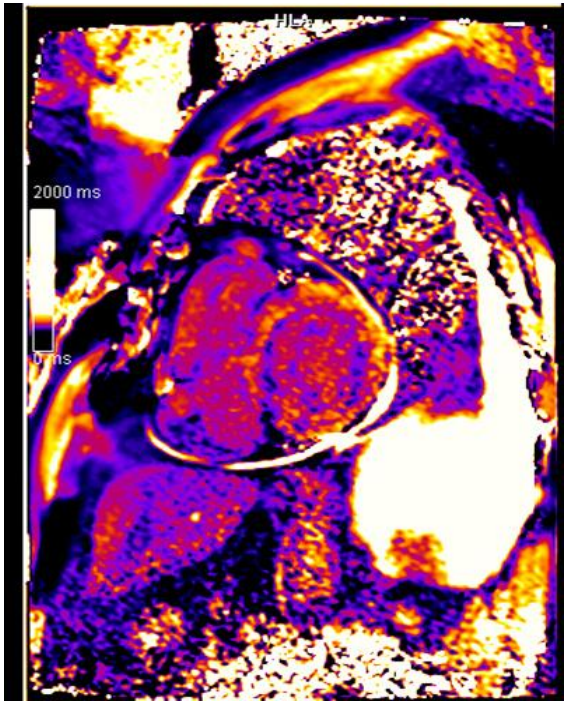
delayed enhanced IR image shows two small thombi at the apex of the left ventricle (arrowhead).

Delayed enhancement MR: Diffuse and bright enhancement of the pericardium.



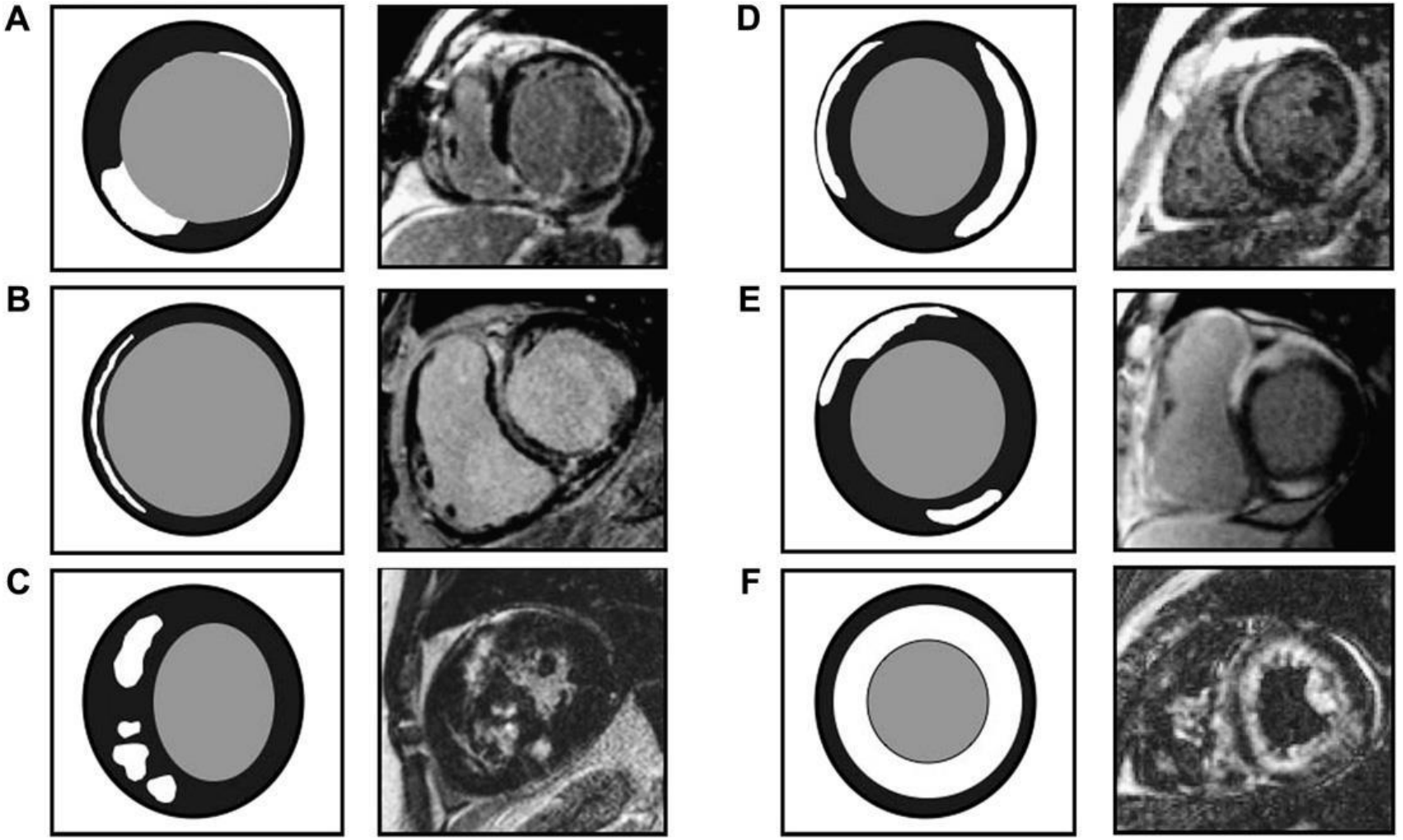




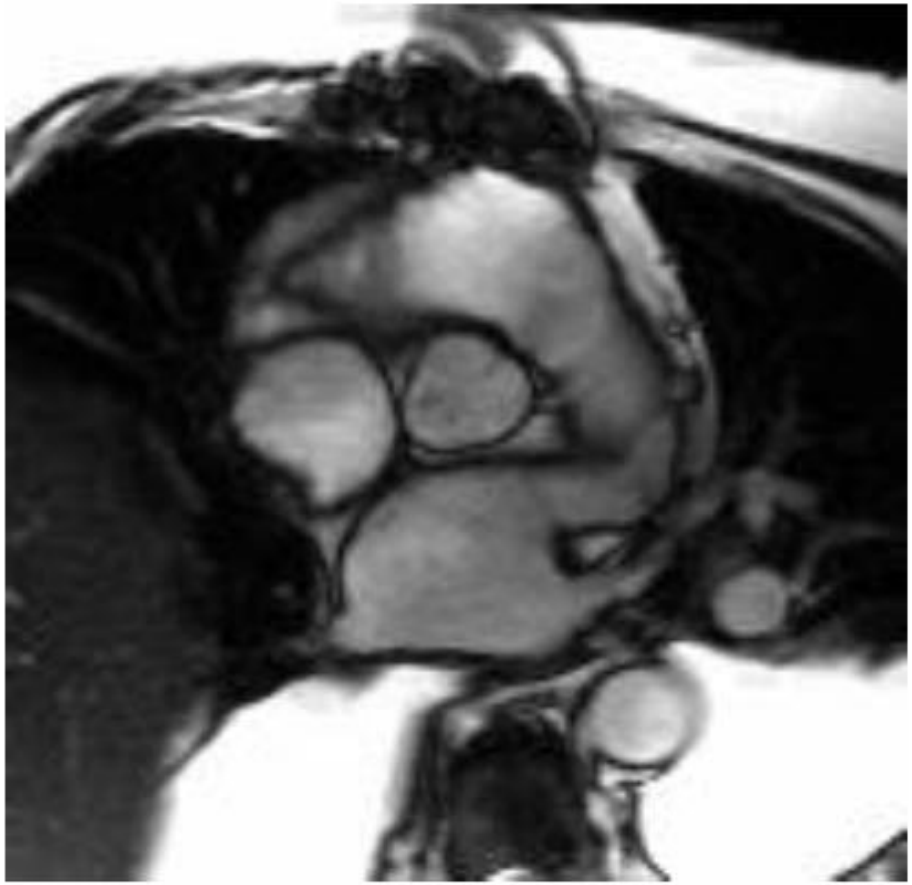


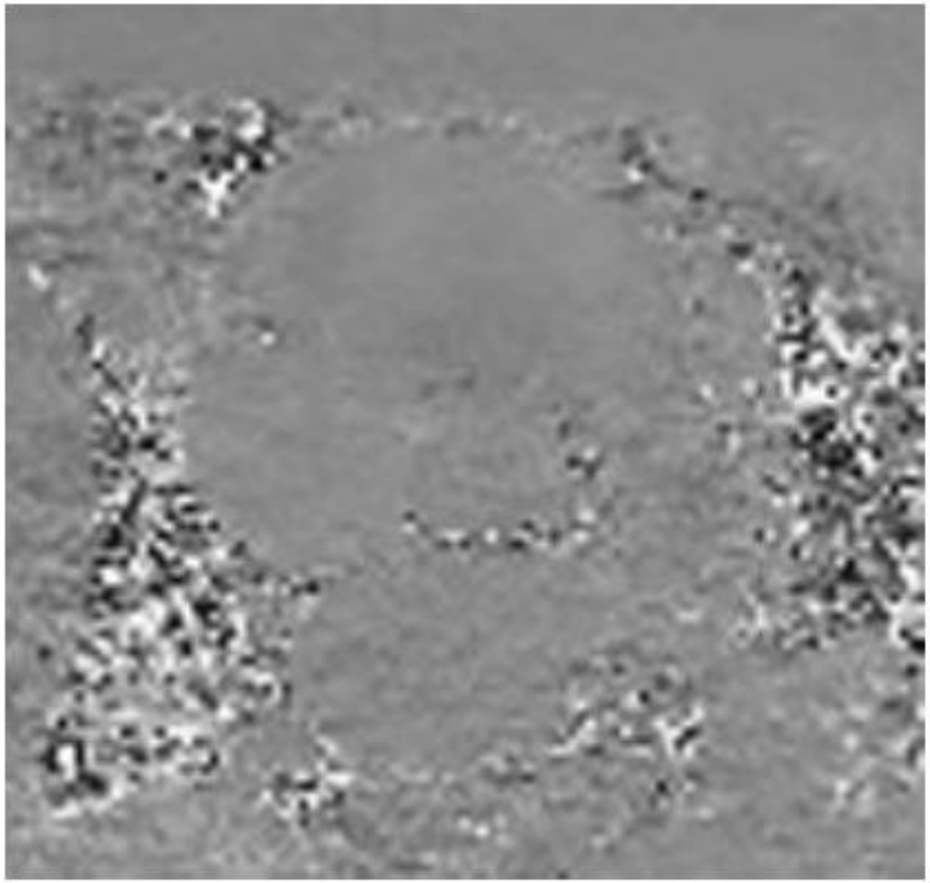
T1 map post-amyloidosis

LGE PATTERNS

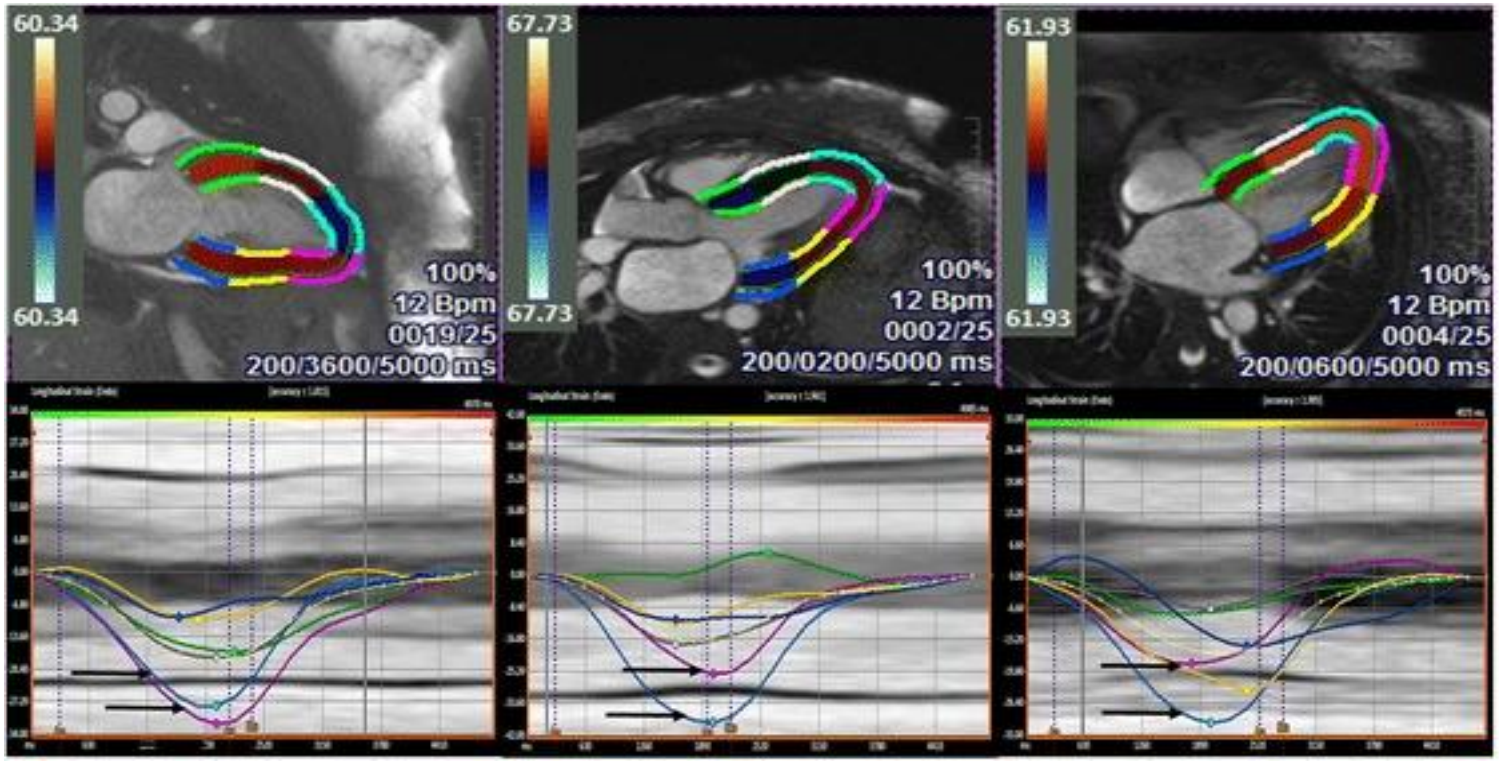


Velocity Flow Mapping





Representative examples of VVI strain analysis and curves in the 2, 3, and 4-chamber long axis views in a patient with cardiac amyloidosis.



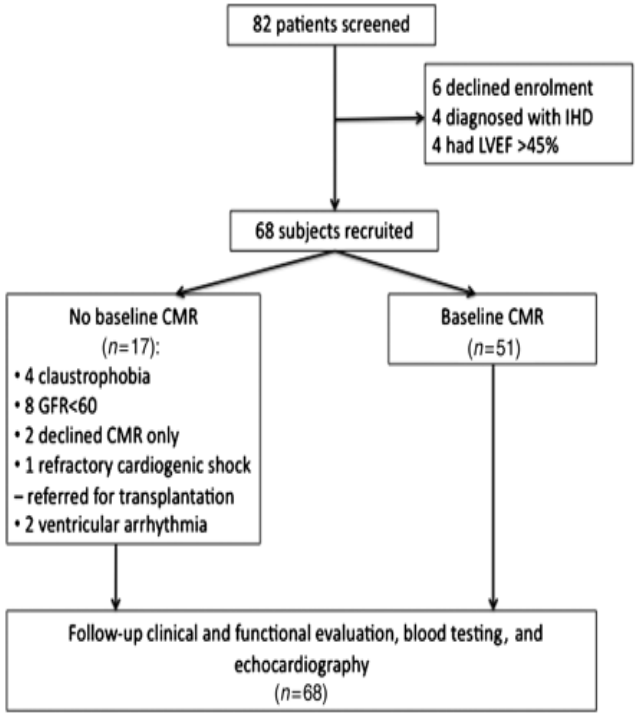
The *lower panel* demonstrates *regional strain curves*, with the *coloured curves* corresponding to the segments in the *upper panel*. In each of the views, the basal segments are represented by the *blue* and *green curves*, the mid-ventricular segments by the *white* and *yellow curves*, and the apical segments by the *turquoise* and *pink curves*. Note the highest regional strain values are consistently seen in the apical segments (highlighted by the *black arrows*) in all three long-axis views.

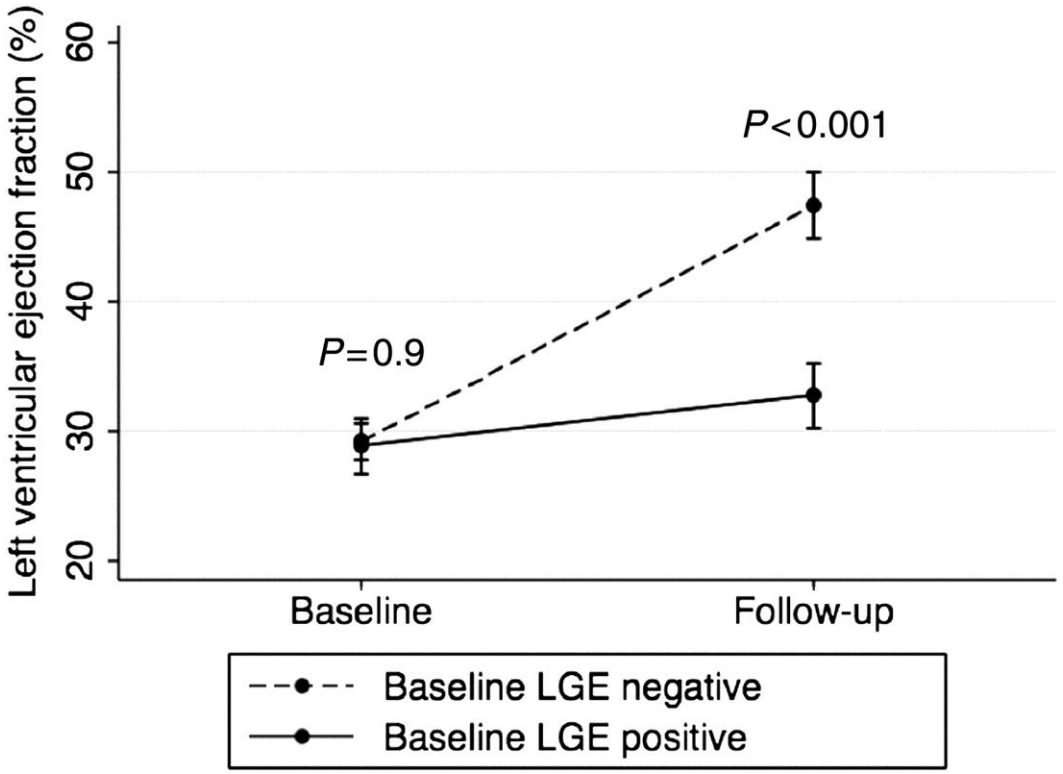
Idiopathic Dilated Cardiomyopathy

- Rule out ischemic cardiomyopathy.
- LGE :
  - Poor response to medical therapy(*)
  - Sudden death and inducible ventricular tachycardia, independent of LV size and function(**).
- The presence and extent of LGE were predictive of all-cause mortality.

- (*) Leong DP, Chakrabarty A, Shipp N, et al: Effects of myocardial fibrosis and ventricular dyssynchrony on response to therapy in new-presentation idiopathic dilated cardiomyopathy: Insights from cardiovascular magnetic resonance and echocardiography. Eur Heart J 33:640, 2012.
- (**) Karamitsos TD, Francis JM, Myerson S, et al: The role of cardiovascular magnetic resonance imaging in heart failure. J Am Coll Cardiol 54:1407, 2009.

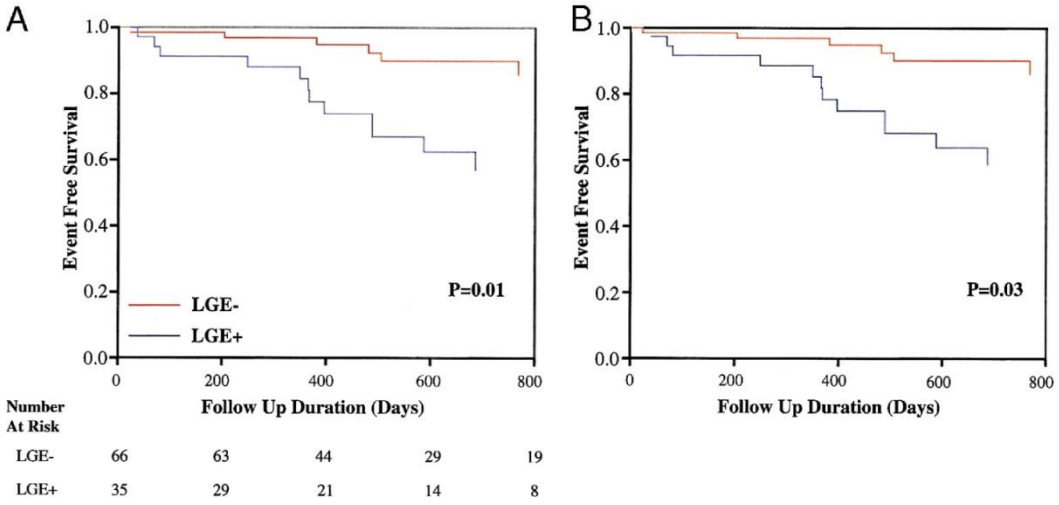
Graph illustrating the influence of baseline late-gadolinium enhancement status on the left ventricular ejection fraction.



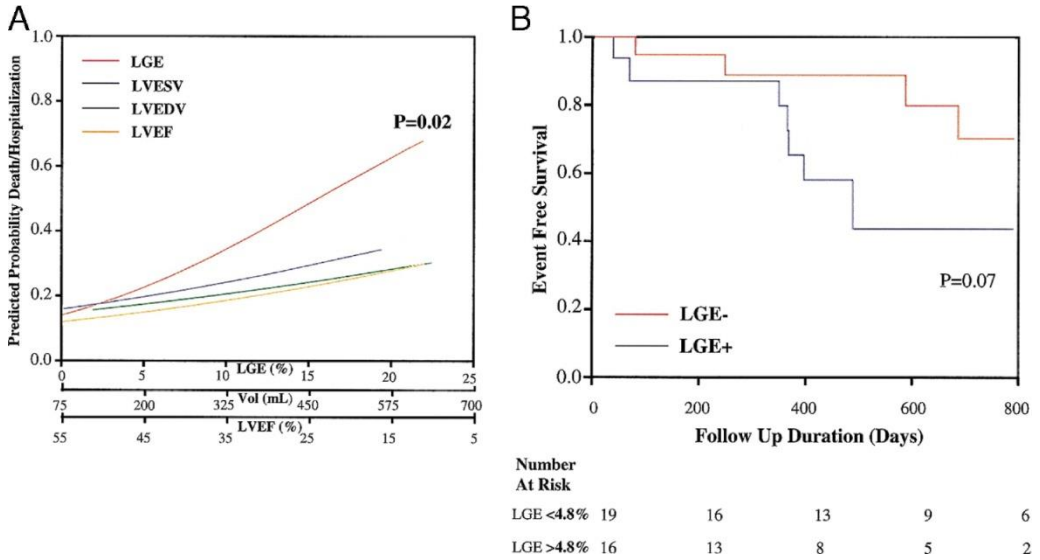


Darryl P. Leong et al. Eur Heart J 2012;33:640-648

European Heart Journal



Kaplan-Meier survival estimates for the secondary end point of sudden cardiac death or sustained ventricular tachycardia. **(B)**Same data adjusted for baseline differences in left ventricular ejection fraction.



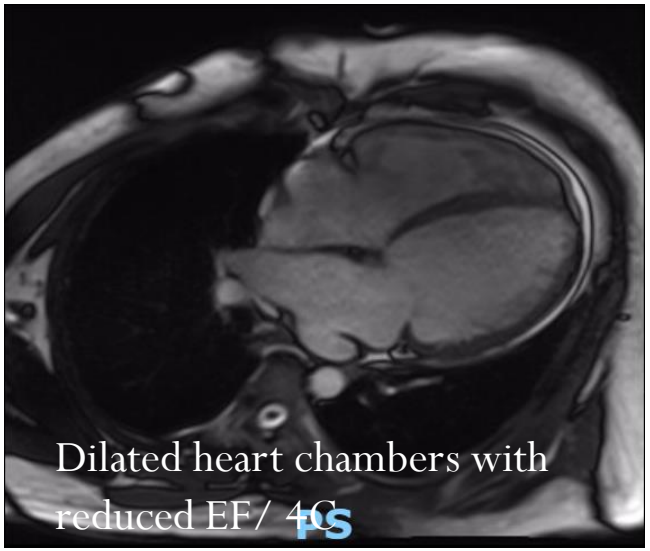
Binary logistic regression analysis comparing the extent of late enhancement (%LGE), left ventricular end-systolic volume (LVESV), left ventricular end-diastolic volume (LVEDV), and left ventricular ejection fraction (LVEF) as predictors of death or hospitalization. There was a strong association between %LGE and outcome, and %LGE was the sole significant predictor of the primary end point (odds ratio 1.12, 95% confidence interval 1.03 to 1.24, p = 0.02)

Ravi G. Assomull . Cardiovascular Magnetic Resonance, Fibrosis, and Prognosis in Dilated Cardiomyopathy

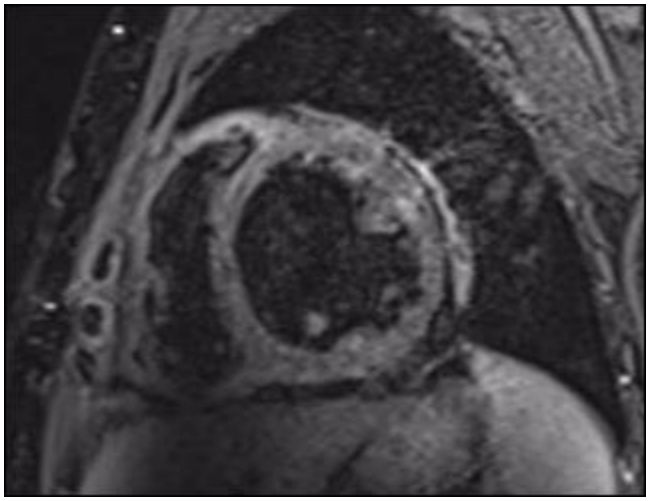
CMR findings in DCMP

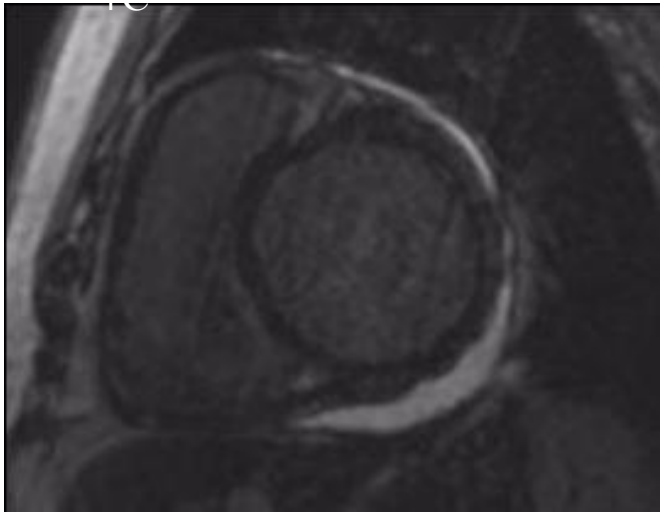
- **Cine:** Dilated LV and/or RV with Reduced LVEF.
- **STIR/ Stress Perfusion:** normal
- **EGE:** Normal or hyposignal intensity within the LV if thrombus.
- **LGE**: Mid-wall septal(commonest) EpicardialDiffuse

CMR in DCMP


Dilated heart chambers with
reduced EF/ 4C


Dilated heart chambers with
reduced contraction/SAX





HYPERTROPHIC CARDIOMYOPATHY

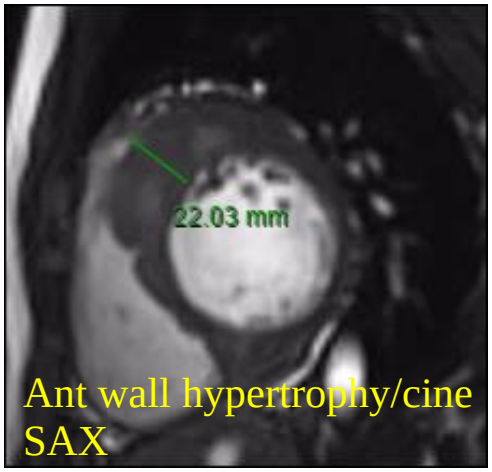
- CINE CARDIAC STRUCTURE/FUNCTION:
  - Localised area of hypertrophy
  - LV apical aneurysm
  - Abnormal motions of hypertrophic wall.
- MYOCARDIAL PERFUSION: abnormal microcirculation -thickened myocardial segments
- LGE IMAGING: fibrosis/ hypertrophied segments
- ASSOCIATED CMR FINDINGS :
  - Phase contrast imaging : Outflow obstruction
  - Systolic anterior motion of mitral leaflet ± mitral regurgitation

HYPERTROPHIC CARDIOMYOPATHY

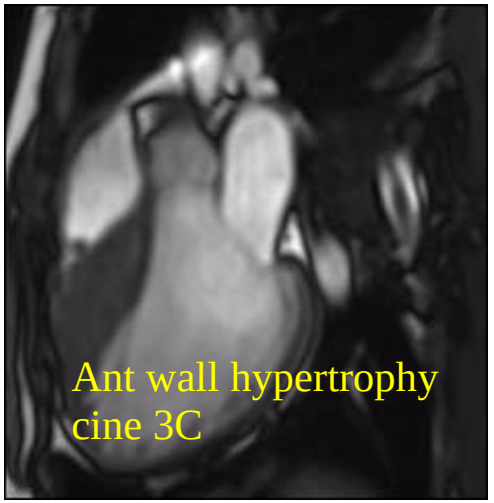
- Diagnosing variant types of HCM, including apical (Yamaguchi's) and lateral wall hypertrophies
- High accuracy in wall thickness measuremenT# prognostic value(*).
- Flow dynamics and dynamic obstruction of the LV outflow tract
- LGE # areas of fibrosis # increased risk of re-entrant tachycardias, ventricular tachycardia, and sudden cardiac death(*)

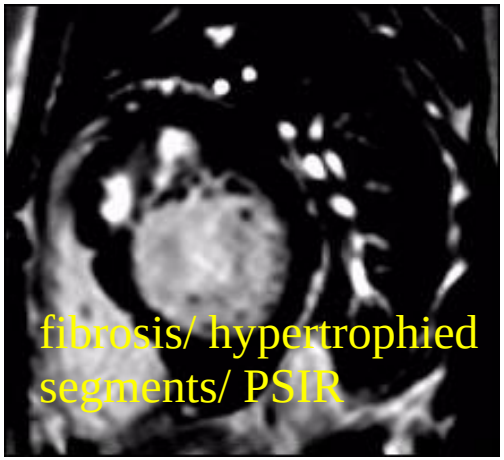
Moon J.C. Toward clinical risk assessment in hypertrophic cardiomyopathy with gadolinium cardiovascular magnetic resonance. J Am Coll Cardiol. 2003;41:1561–1567.

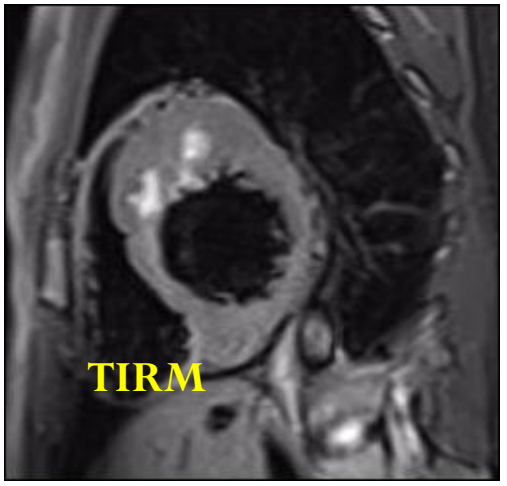
CMR FINDINGS IN HCMP



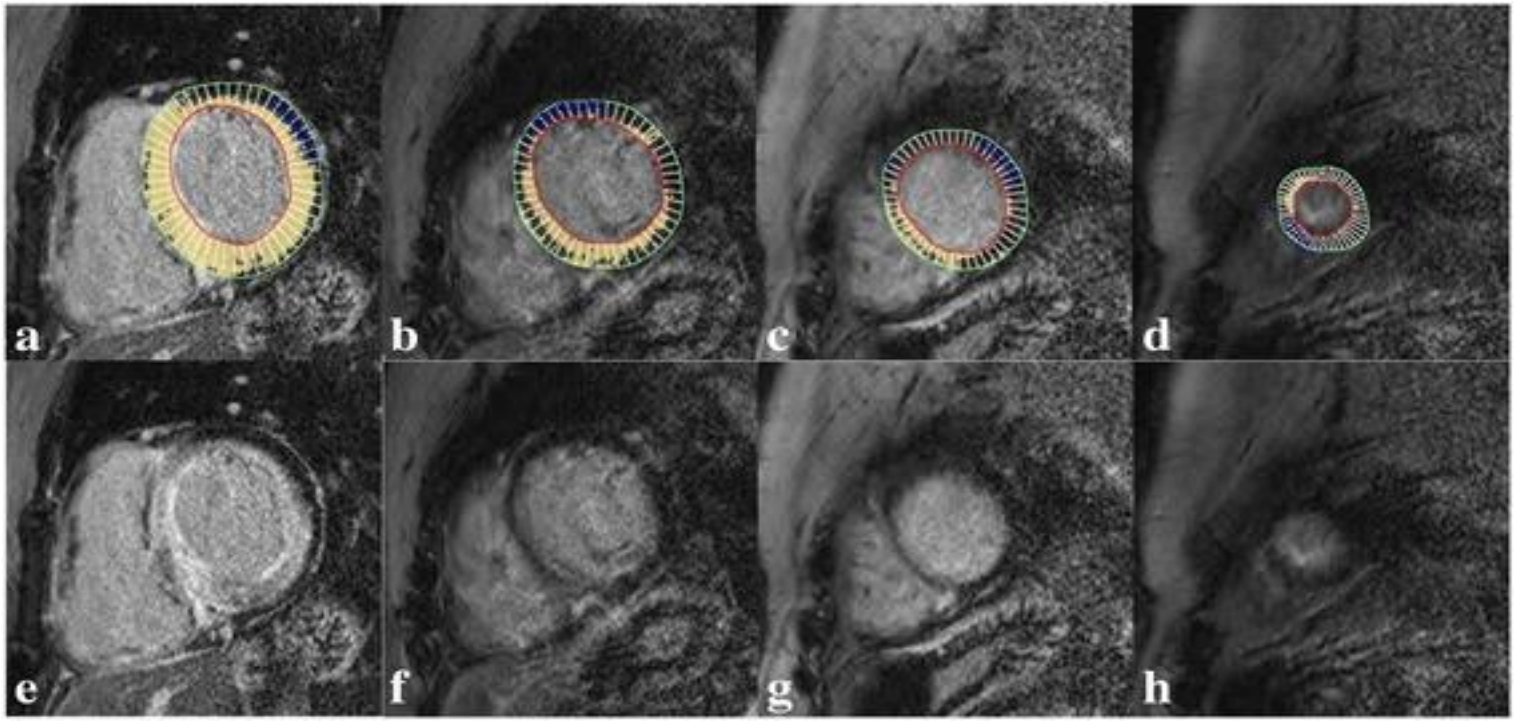
Ant wall hypertrophy/cine SAX


Ant wall hypertrophy cine 3C


fibrosis/ hypertrophied segments/ PSIR


TIRM

Late gadolinium enhancement (LGE) quantification



CoComparison between the contoured (**a**-**d**) and source (**e**-**h**) LGE images in the same slice position to illustrate the quantification method and the resulting difference in the burden of LGE from the basal (**a** and **e**) towards the apical segments (**d** and **h**). For each of the contoured slices (**a**-**d**) the endocardial (*red*) and epicardial contours (*green*) as well as the reference area of non-enhanced myocardium (*blue*) have been defined

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ESC GUIDELINE ON DIAGNOSTIS AND MANAGEMENT OF HYPERTROPHIC CARDIOMYOPATH 2014

Recommendations for cardiovascular magnetic resonance evaluation in hypertrophic cardiomyopathy

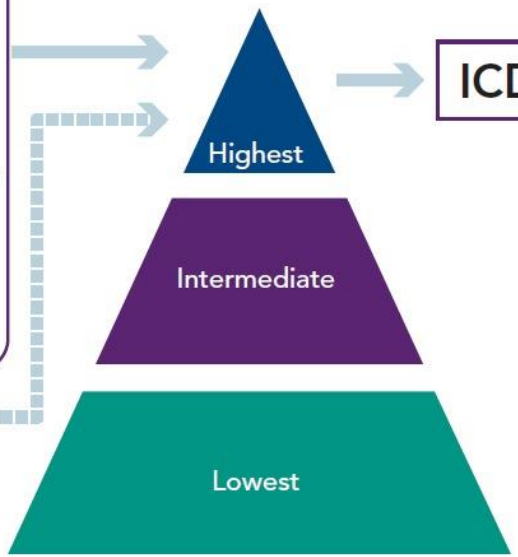
	Class[a]	Level[b]
It is recommended that CMR studies be performed and interpreted by teams experienced in cardiac imaging and in the evaluation of heart muscle disease.	I	C
In the absence of contra-indications, CMR with LGE is recommended in patients with suspected HCM who have inadequate echocardiographic windows, in order to confirm the diagnosis	I	B
In the absence of contra-indications, CMR with LGE should be considered in patients fulfilling diagnostic criteria for HCM, to assess cardiac anatomy, ventricular function, and the presence and extent of myocardial fibrosis.	IIa	B
CMR with LGE imaging should be considered in patients with suspected apical hypertrophy or aneurysm.	IIa	C
CMR with LGE imaging should be considered in patients with suspected cardiac amyloidosis.	IIa	C
CMR with LGE may be considered before septal alcohol ablation or myectomy, to assess the extent and distribution of hypertrophy and myocardial fibrosis.	IIb	C

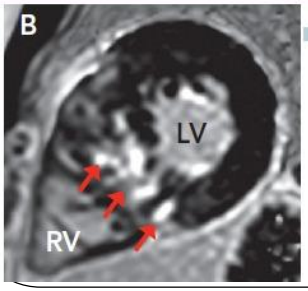
CMR = cardiac magnetic resonance; HCM =

2° Prevention:
Cardiac arrest/sustained VT

1° Prevention:
Familial history of HCM-SD
Unexplained syncope
Multiple-repetitive NSVT
Abnormal exercise BP response
Massive LVH ≥30 mm
*LGE ≥15 % of LV mass**

Rare subgroups:
LV apical aneurysms
End-stage HCM (EF <50 %)



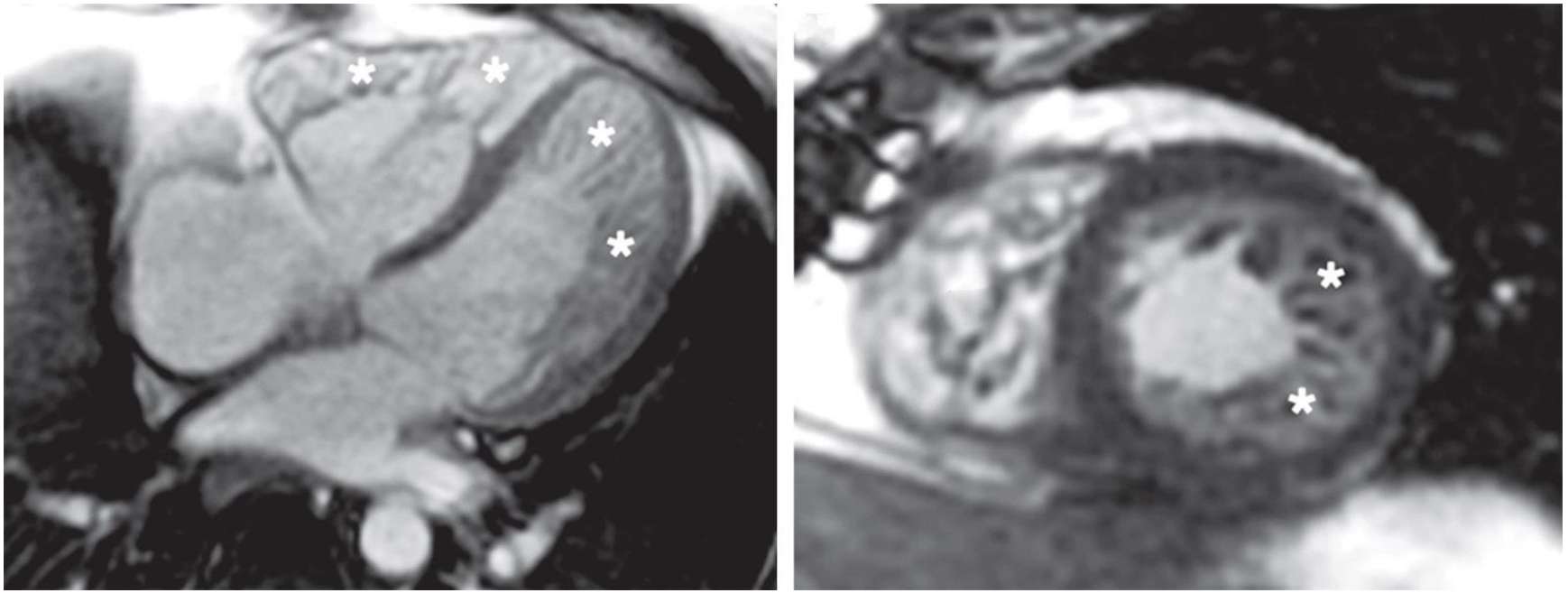


Left ventricular noncompaction (LVNC)

- CMR criteria for diagnosis of LVNC include noncompacted to compacted myocardial thickness ratio of >2.3 (sensitivity, specificity, and positive and negative predictions of 86%, 99%, 75%, and 99%, respectively)
- Trabeculated LV mass >20 percent of global LV mass (sensitivity of 94% and specificity of 94%).

Patel and Kramer JACC: CARDIOVASCULAR IMAGING, VOL. 10, NO. 10, 2017 CMR and Nonischemic Cardiomyopathy

Left ventricular noncompaction (LVNC)



Left ventricular noncompaction. **A, B, Note the heavy trabeculations in the left ventricle (LV) and right ventricle (RV) (*asterisks*), with a ratio of trabeculated myocardium to nontrabeculated myocardium of 5:1. LA = left atrium; RA = right atrium.**
Braunwald's Heart Disease A Textbook of Cardiovascular Medicine, Tenth Edition

Myocarditis

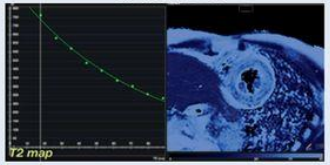
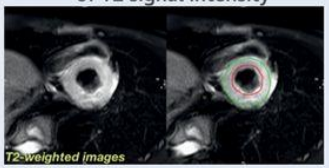
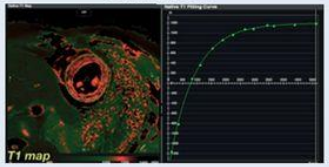
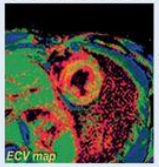
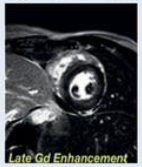
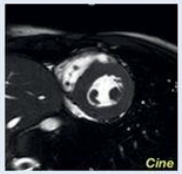
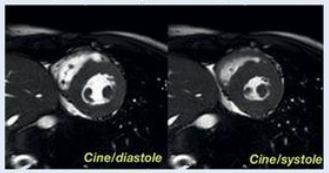
Abdel-Aty H

- CMR targets the three main :
  - Myocardial edema by T2-weighted imaging (ss70%, sp 71%,)
  - Regional hyperemia and capillary leak by early gadolinium enhancement ratio (EGEr) (ss.74%,sp.83%)
  - Myocardial necrosis or fibrosis by LGE imaging (ss59% , sp86%)

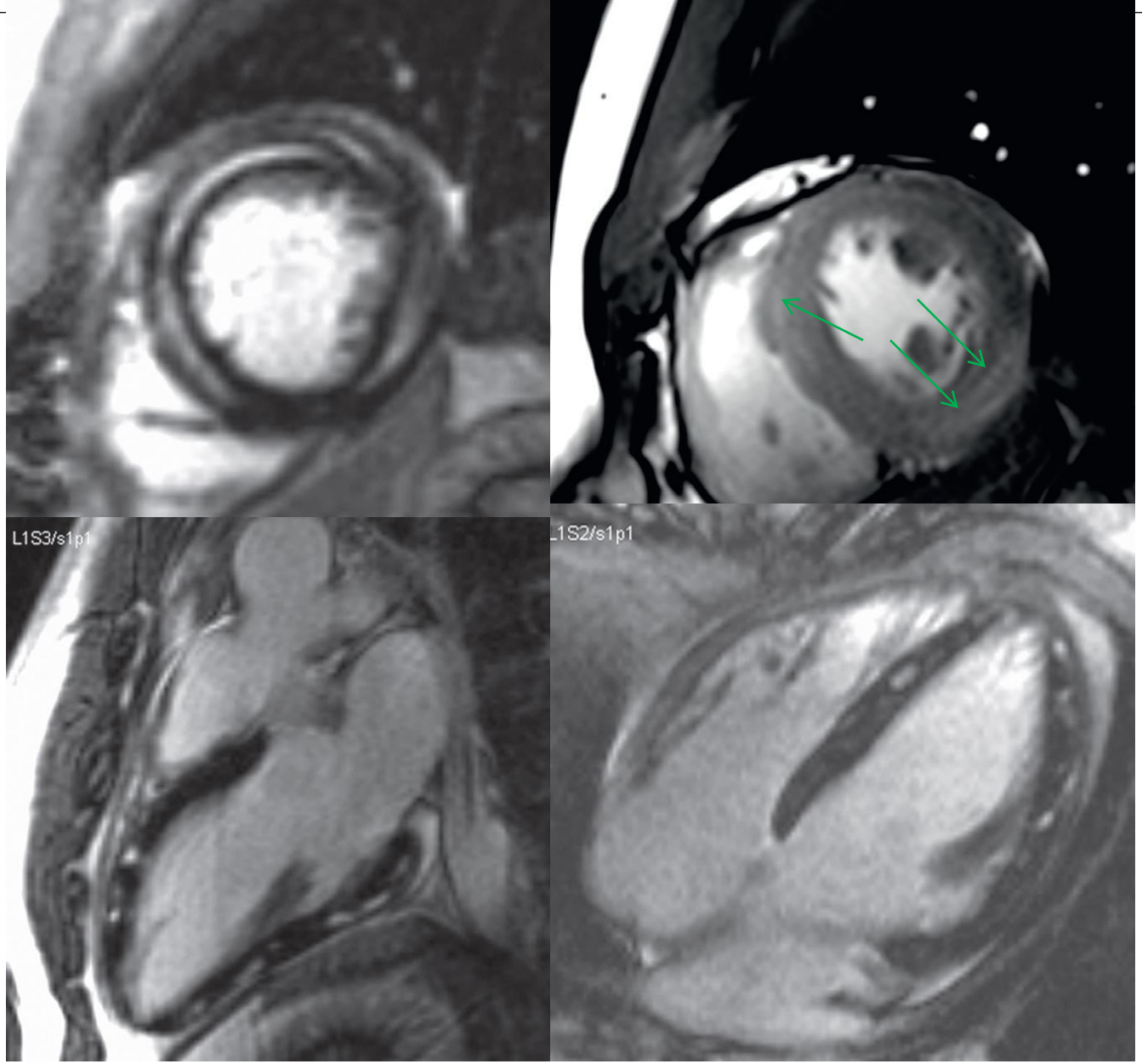
Abdel-Aty H. Diagnostic performance of cardiovascular magnetic resonance in patients with suspected acute myocarditis: comparison of different approaches. J Am Coll Cardiol. 2005

Myocarditis

CENTRAL ILLUSTRATION: Overview of the Updated Lake Louise Criteria

	2018 Lake Louise Criteria	CMR Image Examples
Main Criteria	**Myocardial Edema** (T2-mapping or T2W images)	Regional or global increase of native T2  or Regional or global increase of T2 signal intensity 
	Non-ischemic Myocardial Injury (Abnormal T1, ECV, or LGE)	Regional or global increase of native T1  or Regional or global increase of ECV  or Regional LGE signal increase 
Supportive Criteria	**Pericarditis** (Effusion in cine images or abnormal LGE, T2, or T1)	Pericardial effusion 
	Systolic LV Dysfunction (Regional or global wall motion abnormality)	Regional or global hypokinesis 

Ferreira, V.M. et al. J Am Coll Cardiol. 2018;72(24):3158-76.



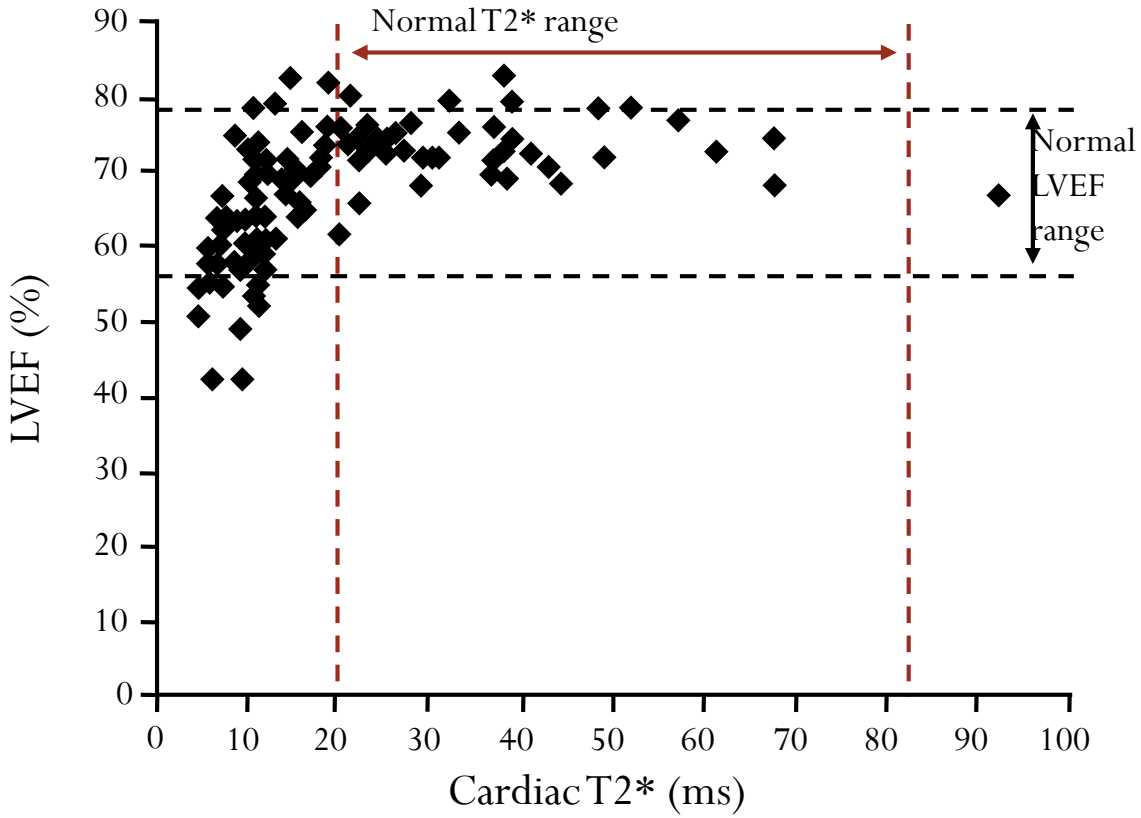
the multiple areas of LGE (**left panels and right lower panel) and diffuse T2 enhancement consistent with edema (right upper panel:** ***green arrows highlighting regions of high signal in this SSFP cine image), strongly suggestive of the diagnosis of acute myocarditis (Braunwald's Heart Disease A Textbook of Cardiovascular Medicine 10th)***

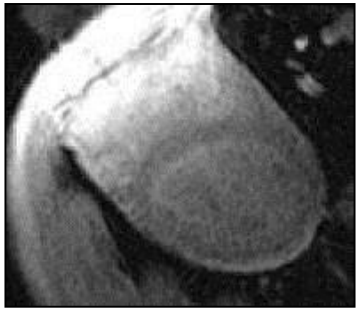
Cardiac Siderosis

- Primary (genetic): hemochromatosis
- Secondary (transfusion dependent) :iron overload.
- T2-star (T2*) technique: ion overload.
- Chelation therapy# cardiomyopathy secondary to iron overload is reversible
- There is no reliable relationship between myocardial T2* and serum ferritin or liver T2*

- *Anderson L. Cardiovascular T2-star (T 2*) magnetic resonance for the early diagnosis of myocardial iron overload. Eur Heart J. 2001*

Cardiac T2*: Relationship with LVEF





Cardiac T2* value of
37 ms in a normal heart

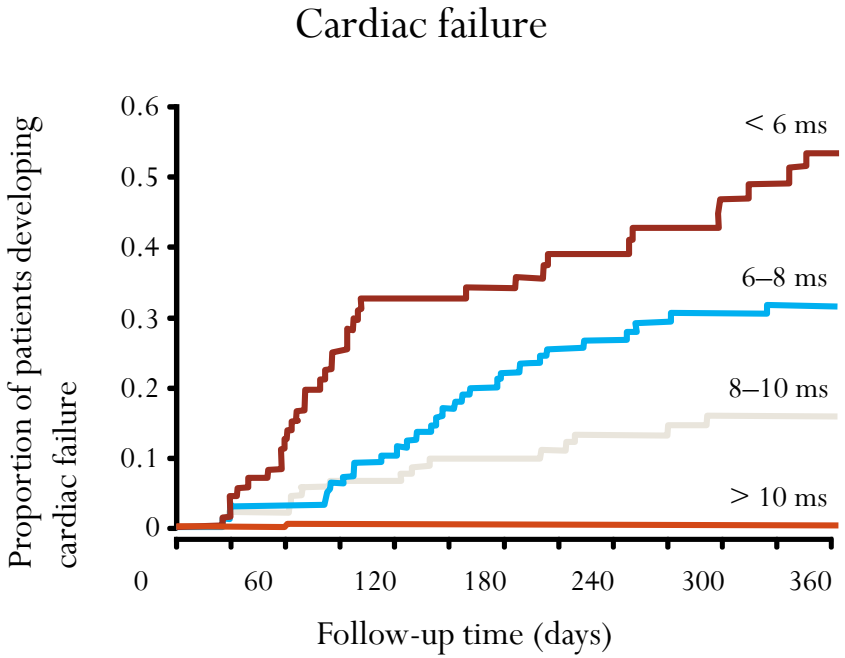
Cardiac T2* value of
4 ms in a significantly
iron-overloaded heart

Myocardial T2* values < 20 ms are associated with a progressive and significant decline in LVEF

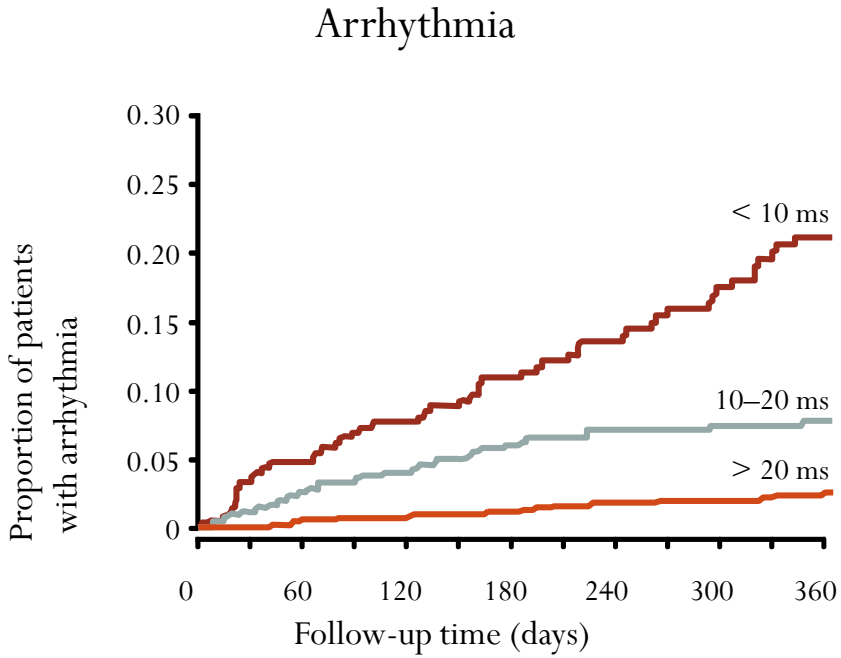
LVEF = left-ventricular ejection fraction.

Anderson LJ, et al. Eur Heart J. 2001;22:2171-9.

Cardiac T2*: Relationship with cardiac failure and arrhythmia



T2* < 10 ms: relative risk 159 ($p < 0.001$)
T2* < 6 ms: relative risk 268 ($p < 0.001$)

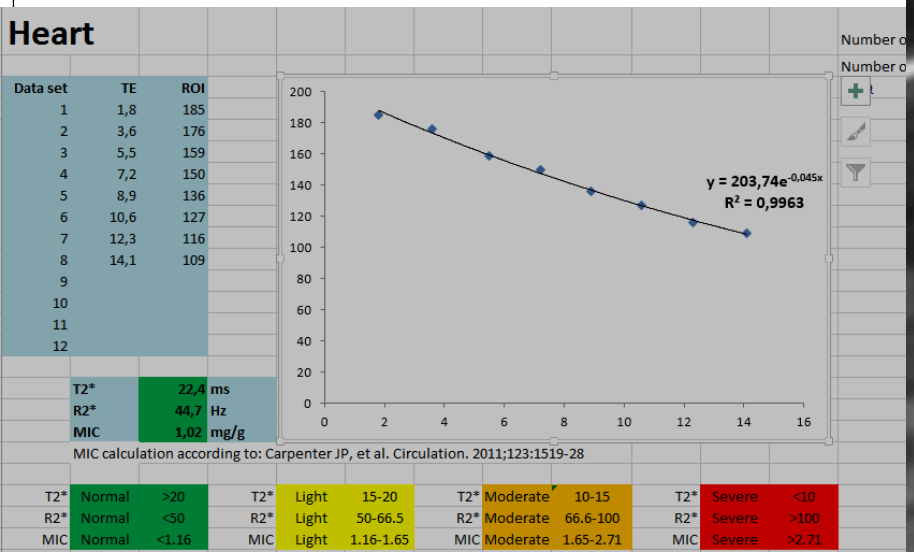


T2* < 20 ms: relative risk 4.6 ($p < 0.001$)
T2* < 6 ms: relative risk 8.65 ($p < 0.001$)

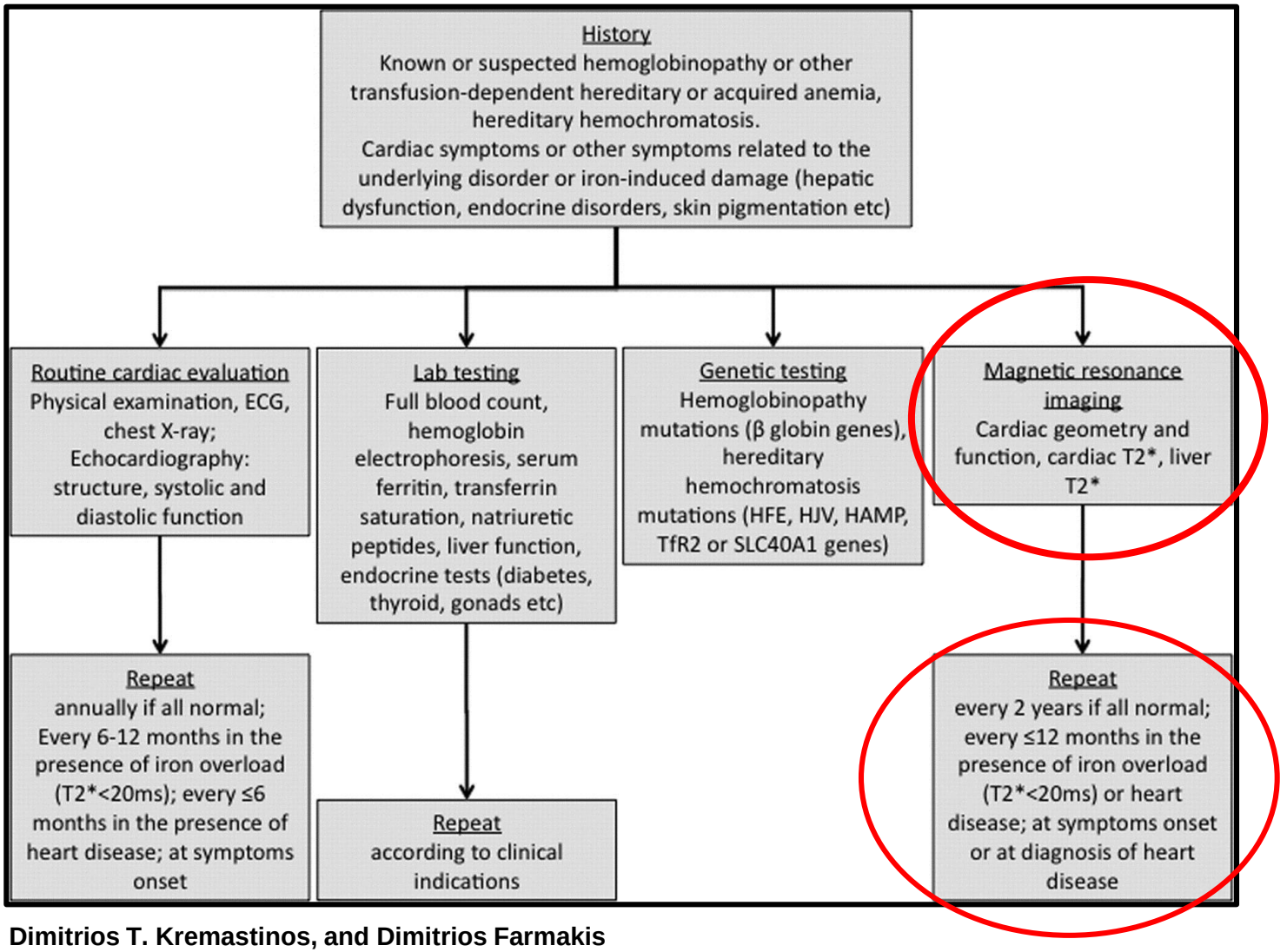
Low myocardial T2* predicts a high risk of developing cardiac failure and arrhythmia

Kirk P, et al. *Circulation*. 2009;120:1961-8.

Manually	Post-processing software
• Excel spreadsheet	• ThalassaemiaTools (CMRtools) • cmr^{42} • FerriScan • MRmap • MATLAB



A proposed algorithm for the diagnostic evaluation and follow-up of patients with known or suspected iron overload cardiomyopathy or at risk for iron overload cardiomyopathy.



Dimitrios T. Kremastinos, and Dimitrios Farmakis
Circulation. 2011;124:2253-2263

American Heart Association.

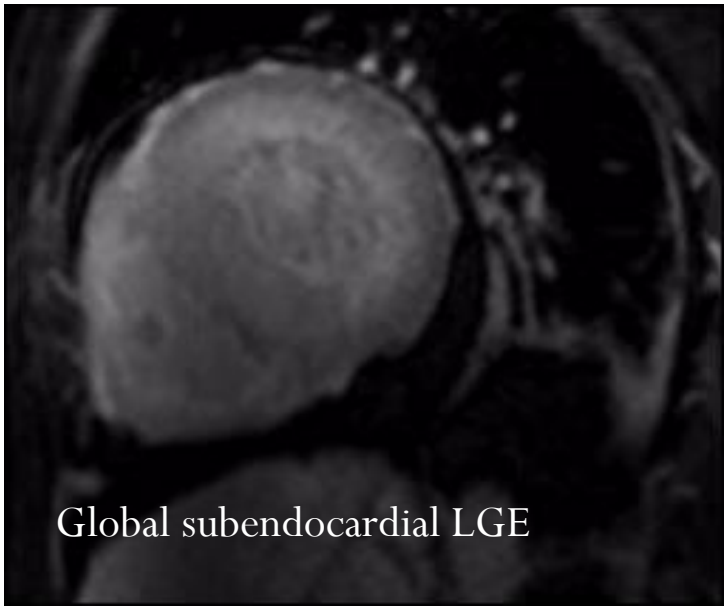
Amyloidosis

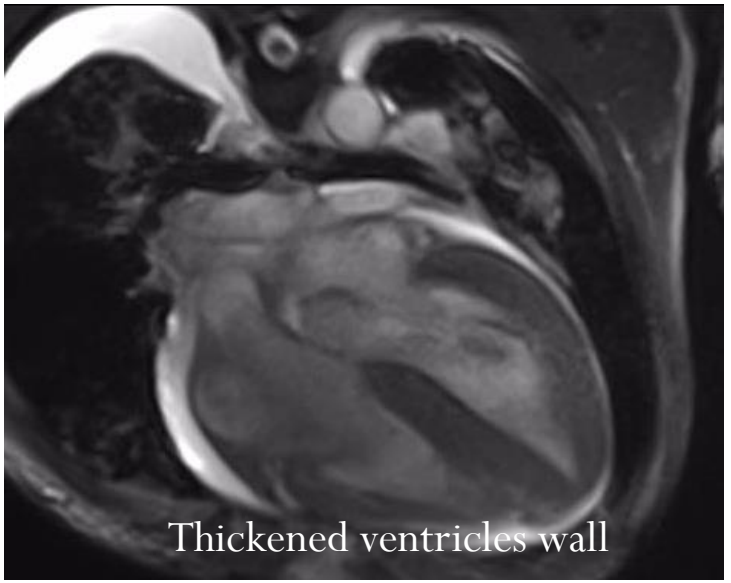
- CMR with LGE has been shown to have greater sensitivity and specificity than TTE.
- Cardiac amyloidosis appears as global subendocardial myocardial hyperenhancement on LGE.
- CMR with LGE has been reported to have sensitivity, specificity, positive predictive, and negative predictive values of between 86–88%, 86–90%, 88–95%, and 67–90%.
- *Ruberg F.L. Diagnostic and prognostic utility of cardiovascular magnetic resonance imaging in light-chain cardiac amyloidosis. Am J Cardiol. 2009;103:544–549.*

CMR findings

- **Restrictive LV pattern**
- **Global LV hypertrophy**
- **LGE**
  - Difficult to NULL
  - **Sub-endocardial distribution/** patchy subendocardial or transmural
  - Cardiac involvement without any LGE is rare
- **Atrial septum hypertrophy of >6mm** (20% of cases)
- **Pericardial** and **pleural effusion**

CMR IN AMYLOIDOSIS





CMR of 36yo man admitted to UMC because of new onset of heart failure.
Left: Diffuse circumferential LGE and difficulty in finding the right inversion time in "nulling" normal myocardium signal during LGE imaging.
Right: Cine imaging Thickened ventricles wall.

CMR-based differentiation of amyloidosis types

	AL amyloidosis	ATTR amyloidosis
LV Mass	• Mildly increased < 100 g/m2	• Markedly increased > 100 g/m2
Septum thickness	Septum AL < Septum ATTR	
LGE	• Less extensive LGE • Often (global)	• More extensive LGE • Often more diffuse and
Native T1	Native $T1_{AL}$ (> 1050 - 1150 ms) > Native $T1_{ATTR}$	
ECV	ECV_{AL} < ECV_{ATTR} (>0.40)	
Therapy	• Chemotherapy	• Novel TTR-specific treatment (Phase III)
Prognosis	• Worse (despite less extensive LGE)	• Better (despite more extensive LGE)

- Dungu JN, Valencia O, Pinney JH, et al. *CMR-based differentiation of AL and ATTR cardiac amyloidosis.* JACC Cardiovasc Imaging. 2014 Feb;7(2):133-42.

CONCLUSION

- Cardiomyopathy is often suspected on the basis of symptoms, an associated abnormal ECG and echocardiographic findings.
- CMR with LGE, T1 mapping, ECV, T2 mapping, and T2* imaging provides important insights into the underlying etiology of cardiomyopathy.
- Allowing pre-clinical detection of disease process and novel parameters for risk stratification.


Thank you for your attention