Cardiomyopathies

- Clinical practice & unmet needs -

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COI: speakers fees & advisory boards Bayer AG

USZ Universitäts Spital Zürich

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University of Zurich[UZH]

Cardiovascular Imaging @ USZ

Scientific

Institute of Diagnostic and Interventional Radiology

University Heart Center Zurich

Improved Diagnosis

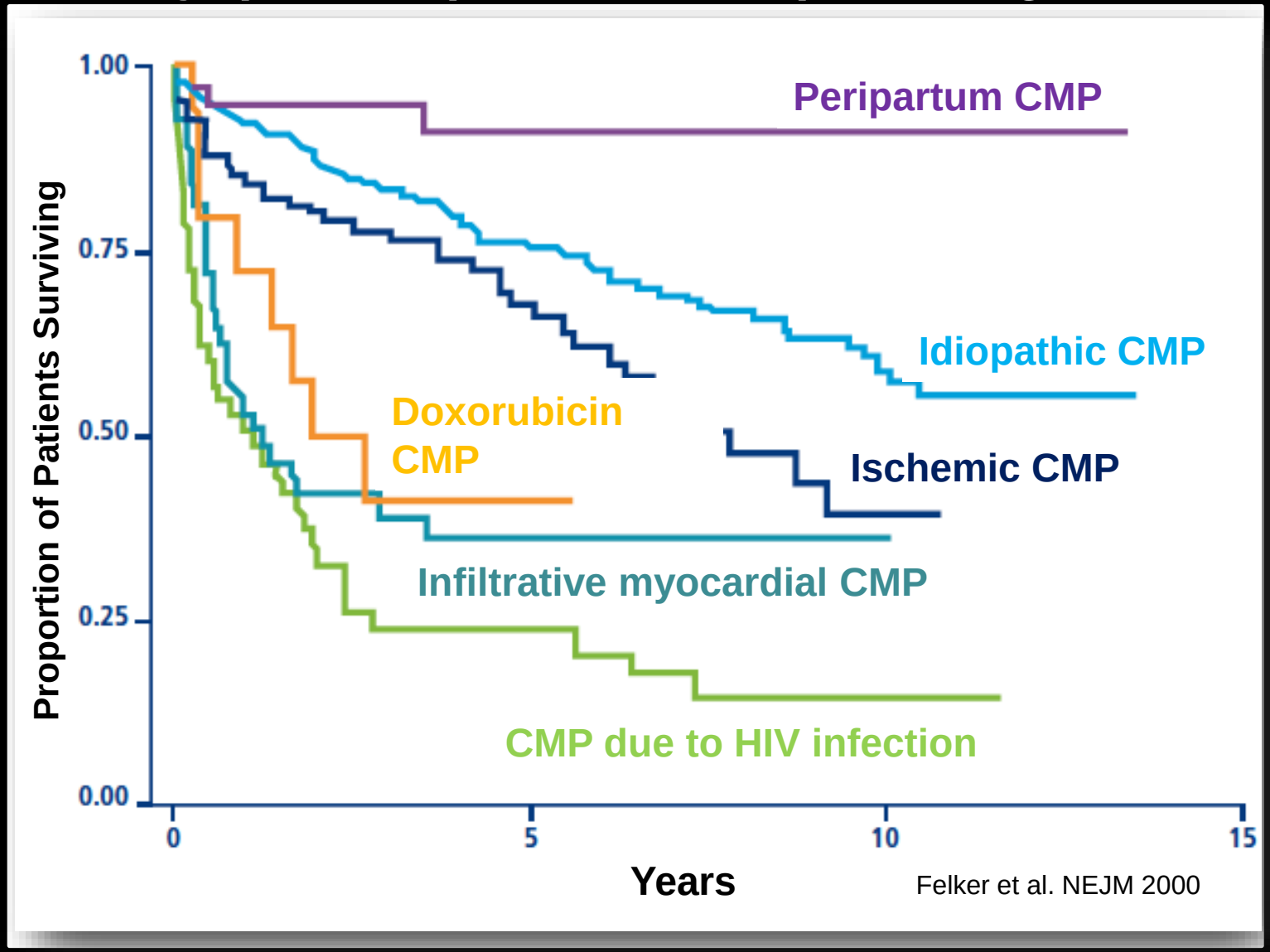
Enhar

eted treatment

al Research

Cardiomyopathies

- Classification -
- Cardiomyopathies (non-ischemic) have high mortality -



ESC Guidelines

- Cardiovascular MRI -

- Goldstandard for the assessment of **volumes**, **mass** and **EF**
  - High accuracy and reproducibility

- Preferred imaging method to assess **myocardial fibrosis** using
  - late gadolinium enhancement (LGE) – *focal fibrosis*
  - T1 mapping – *diffuse interstitial fibrosis*

Recommendations	Class[a]	Level[b]
CMR		
CMR is recommended for the assessment of myocardial structure and function in those with poor echocardiogram acoustic windows.	I	C
CMR is recommended for the characterization of myocardial tissue in suspected infiltrative disease, Fabry disease, inflammatory disease (myocarditis), LV non-compaction, amyloid, sarcoidosis, iron overload/haemochromatosis.	I	C
CMR with LGE should be considered in DCM to distinguish between ischaemic and non-ischaemic myocardial damage.	IIa	C

McDonagh TH et. al. EHJ 2021

Diagnostic Modules

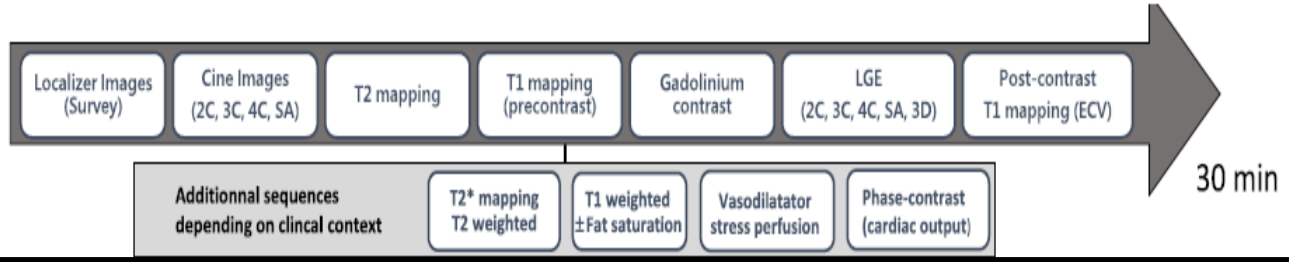
- Cardiovascular MRI -

ESC European Society of Cardiology
European Heart Journal - Cardiovascular Imaging (2022) **23**, 587–589
https://doi.org/10.1093/ehjci/jeac051

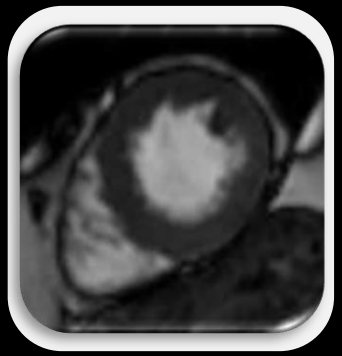
HOW TO

How to evaluate cardiomyopathies by cardiovascular magnetic resonance parametric mapping and late gadolinium enhancement

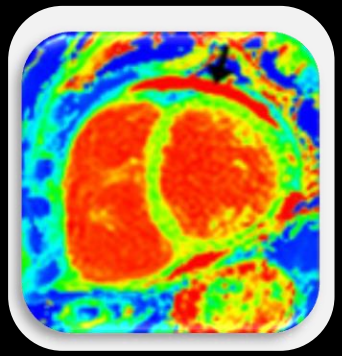
Nassiba Menghoum[1], Jacqueline L. Vos[2], Anne-Catherine Pouleur[1], Robin Nijveldt[2], and Bernhard L. Gerber [1]*



Function/Strain

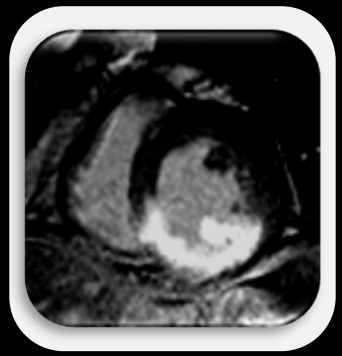


Tissue



Ischemia

Fibrosis/Scar



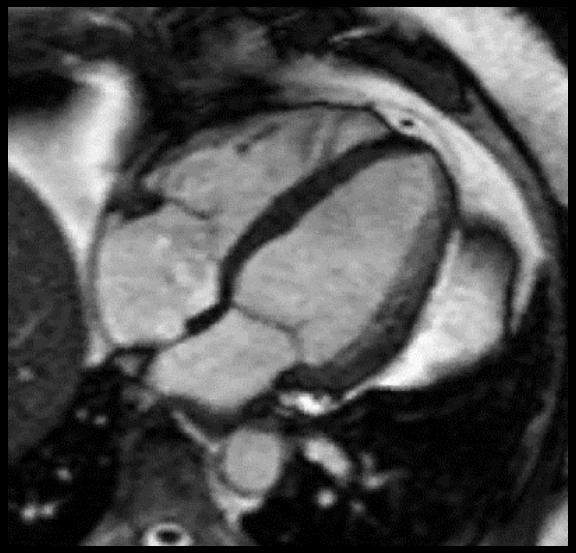
Cardiomyopathy
- Diagnosis and Etiology-

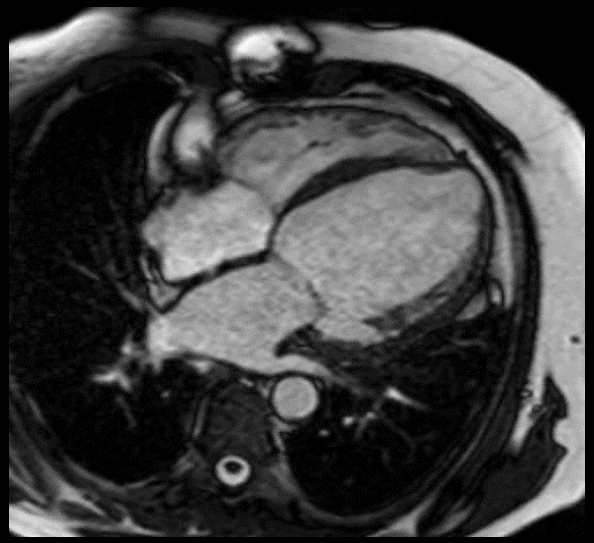
Left & right ventricle

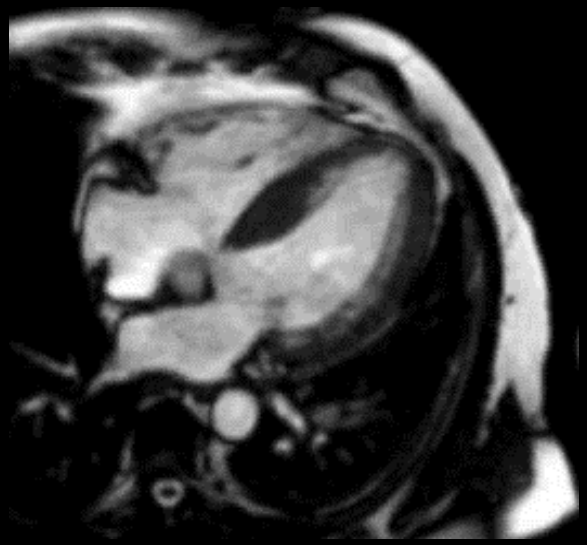
normal

dilatation

hypertrophy





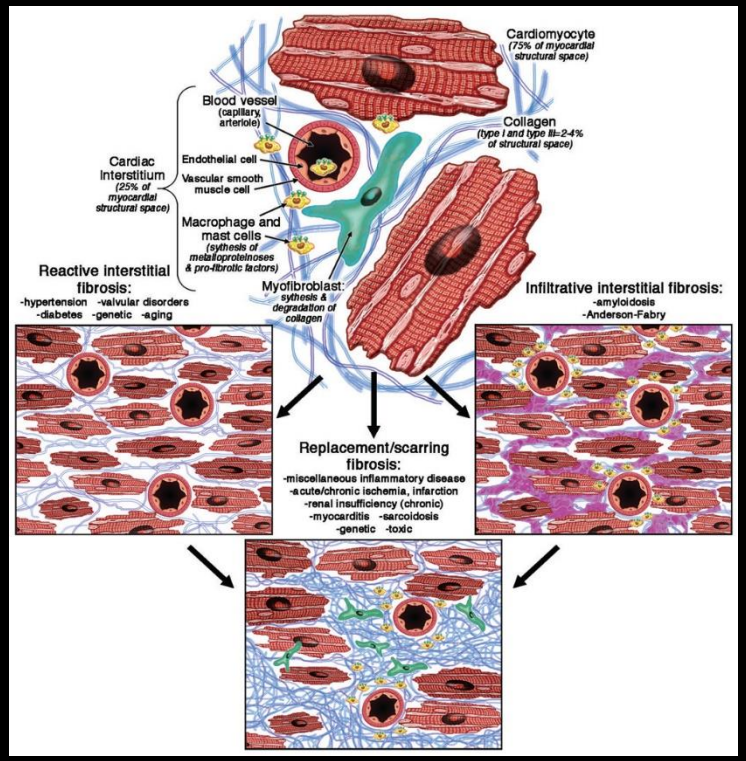


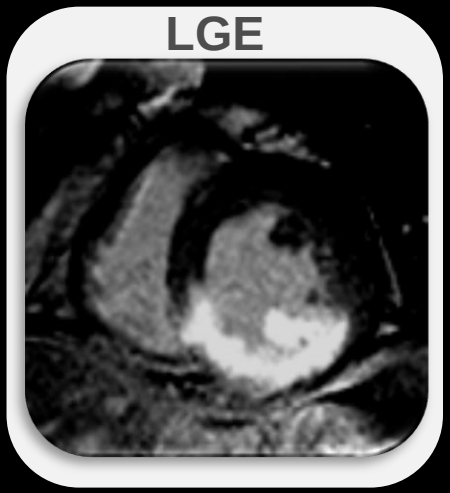
Diagnostic Modules

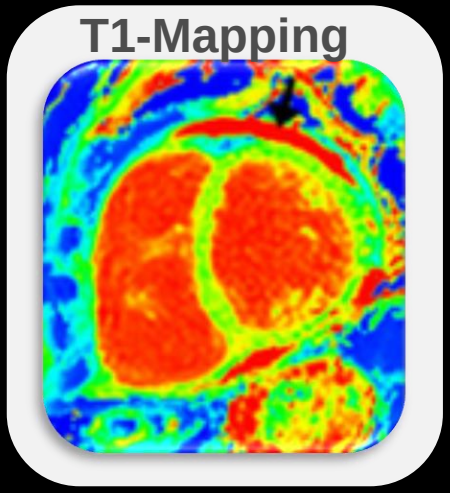
- Fibrosis/Scar -

Types of fibrosis:

- **Reactive interstitial**
  - *AS, HT, Age*
- **Replacement/scarring**
  - *MI, Myocarditis, Toxic, Sarcoid*
- **Infiltrative interstitial**
  - *Amyloid, Fabry*



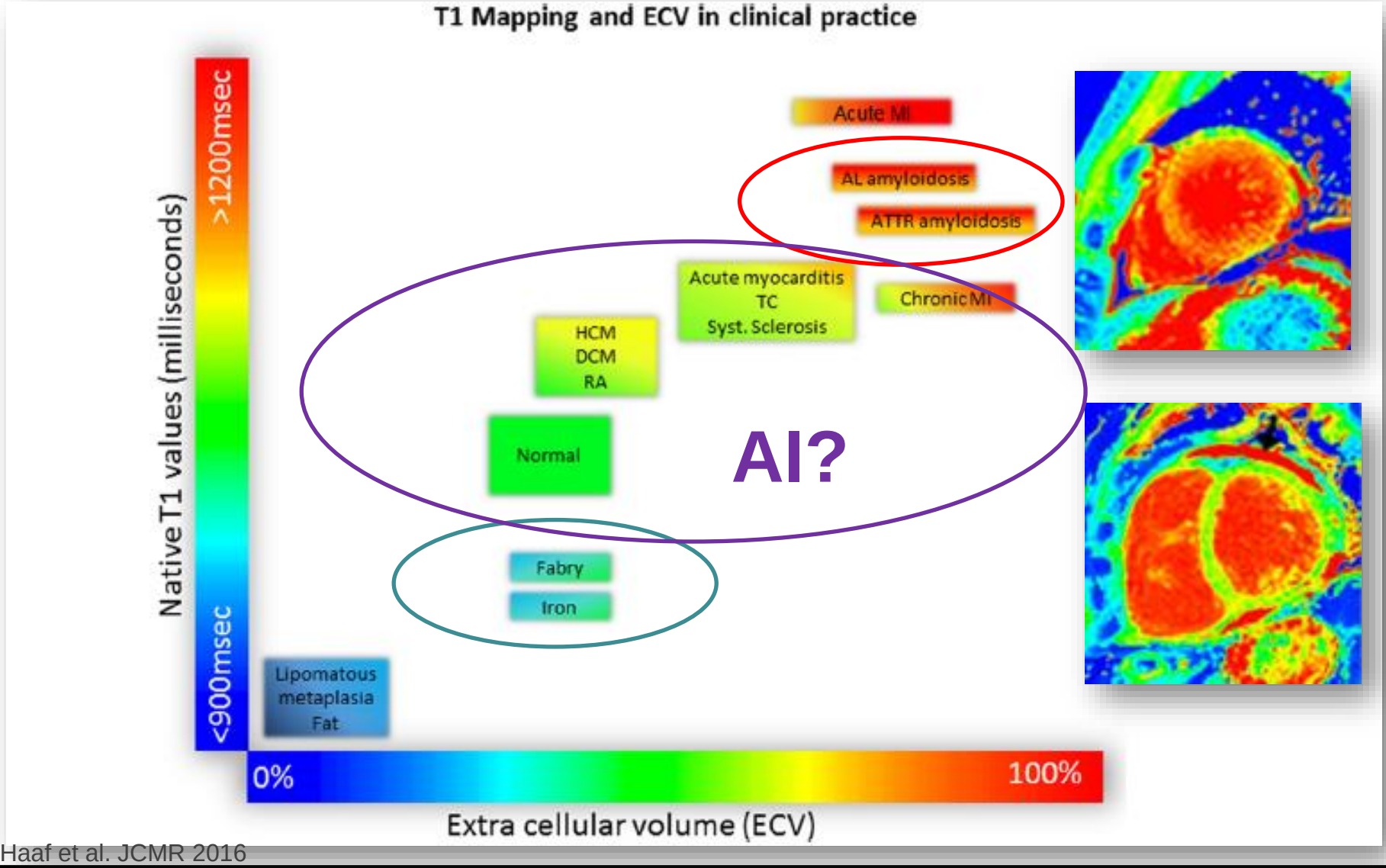




Mewton et al. JACC 2011

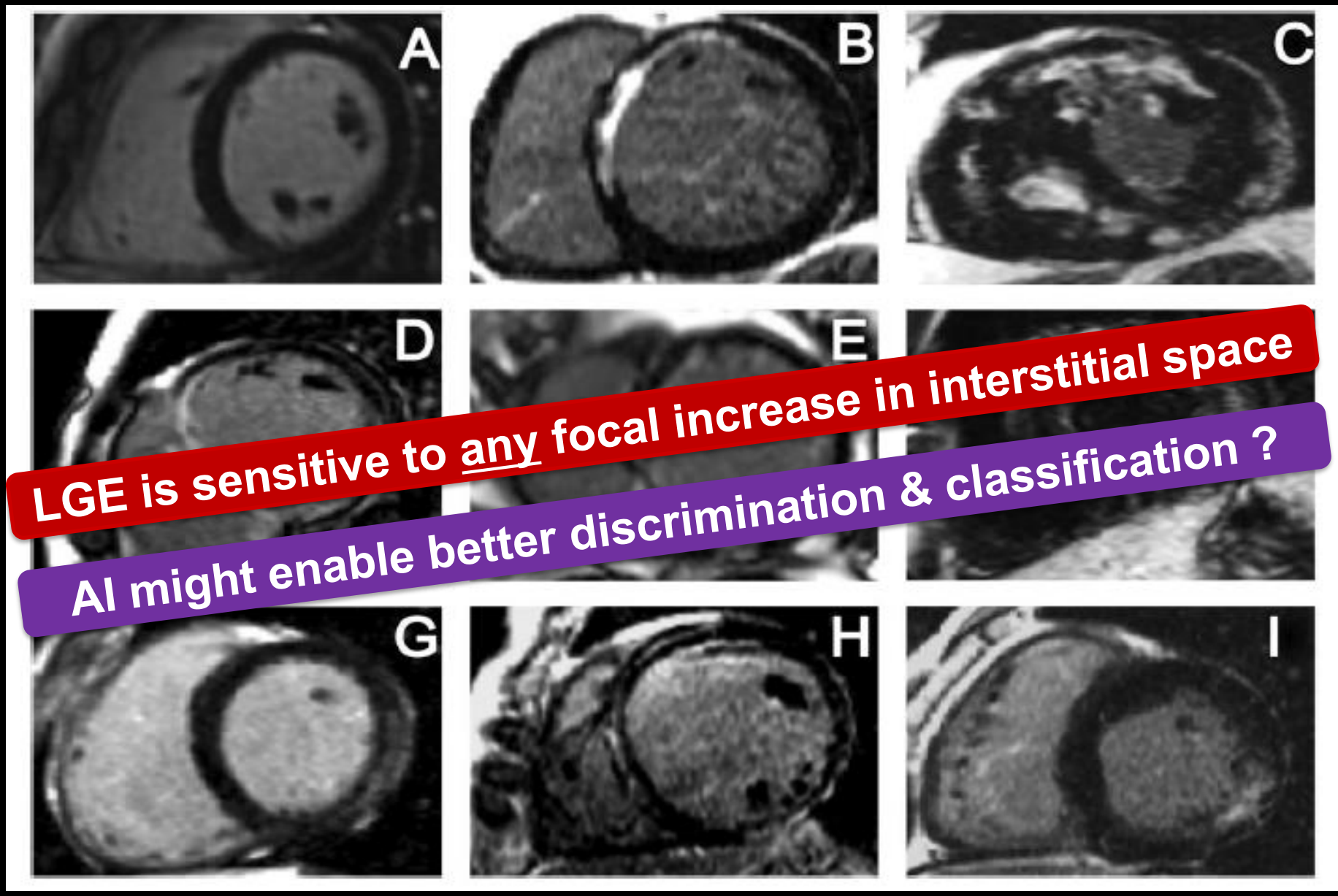
Diffuse fibrosis

- native T1 Mapping (without contrast agent) -



Haaf et al. JCMR 2016

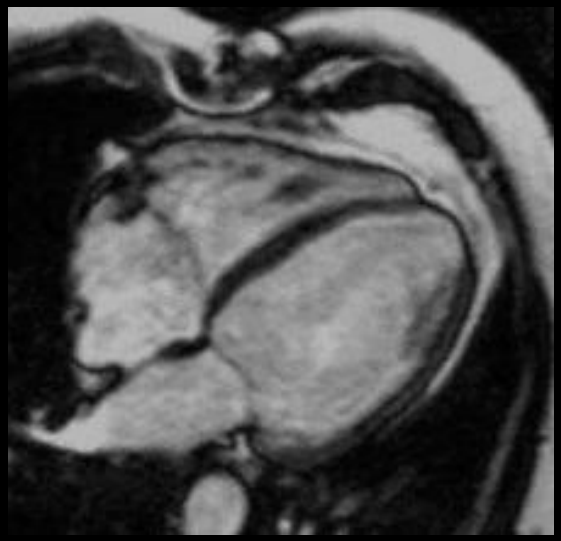
Late Gadolinium Enhancement (LGE)

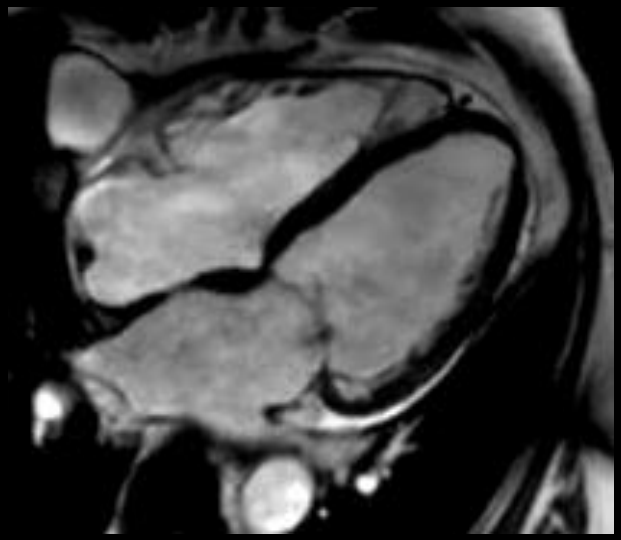


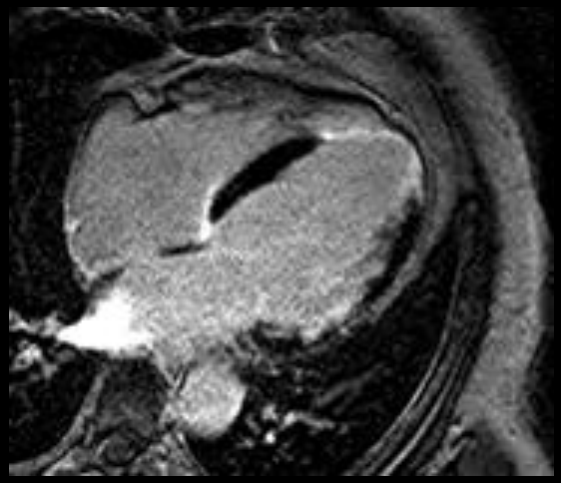
Mahrholdt H, et al. EHJ 2005

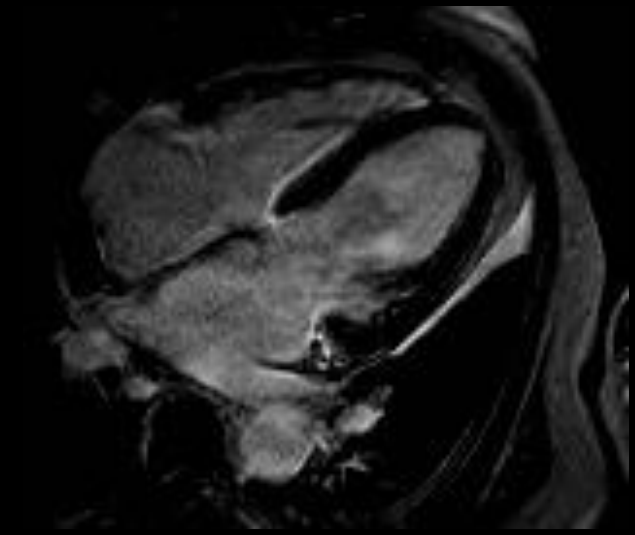
Chronic Left ventricular dysfunction

- Ischemic vs. Non-Ischemic-





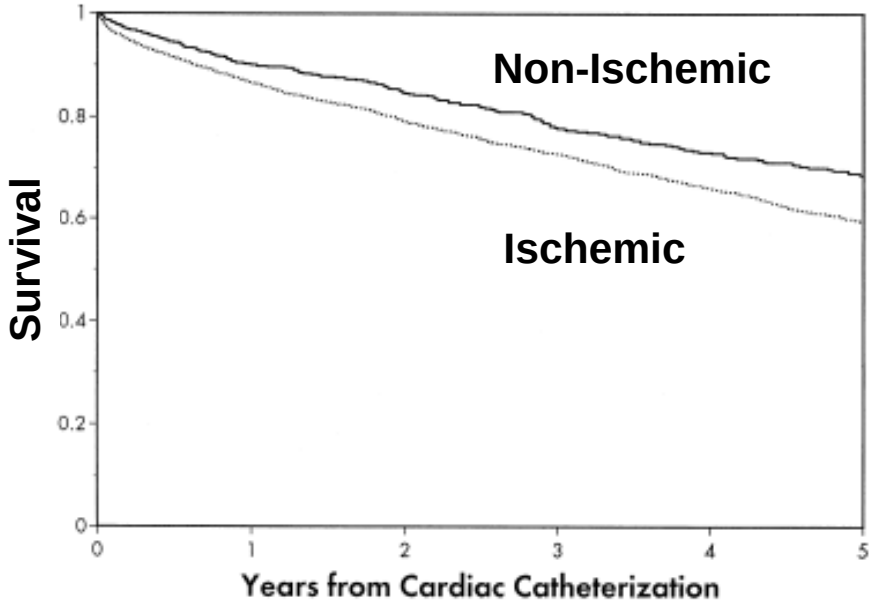




Prognosis Etiology

- Ischemic vs. Non-Ischemic -

Figure 3. Adjusted Kaplan-Meier survival estimates for patients with nonischemic (**solid line**) and ischemic (**dashed line**) cardiomyopathy ($p < 0.0001$).



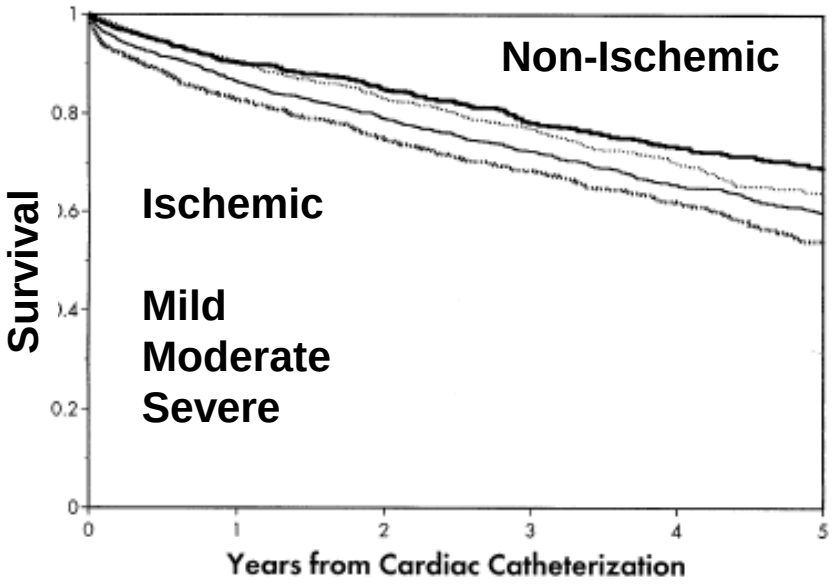


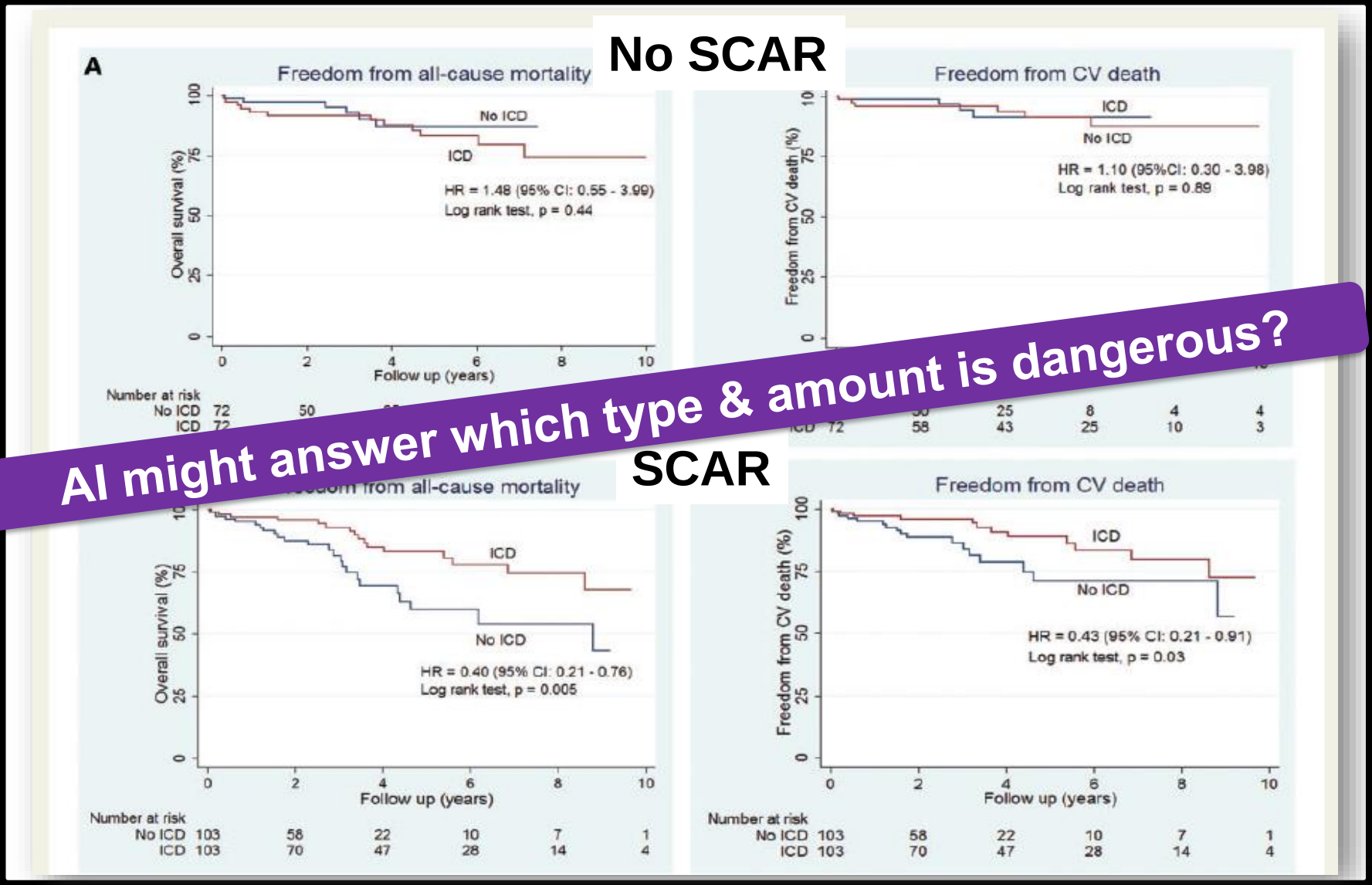
Figure 4. Adjusted Kaplan-Meier survival estimates for patients with nonischemic cardiomyopathy (**heavy solid line**), ischemic cardiomyopathy with mild CAD (**light dashed line**), ischemic cardiomyopathy with moderate CAD (**light solid line**) and ischemic cardiomyopathy with severe CAD (**heavy dashed line**).

N= 3787, LVEF < 40%, 11y follow-up

Bart et al. JACC 1997

Prognosis Non-Ischemic CMP

- Scar & ICD -

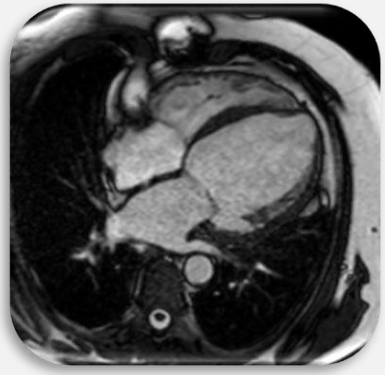


N = 452, LVEF ≤35%, NYHA II-III & Guideline Indication for ICD

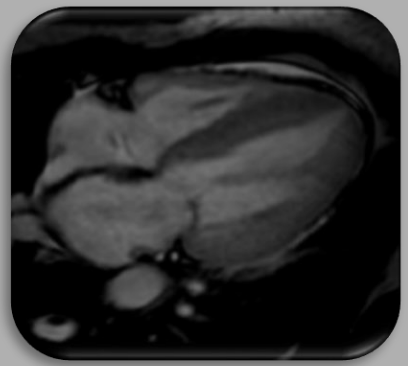
Gutmann et al. Eur Heart J 2019

Non-Ischemic Cardiomyopathies

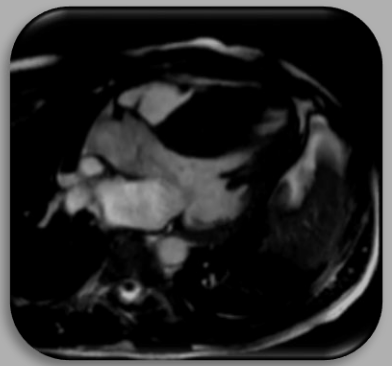
Dilated



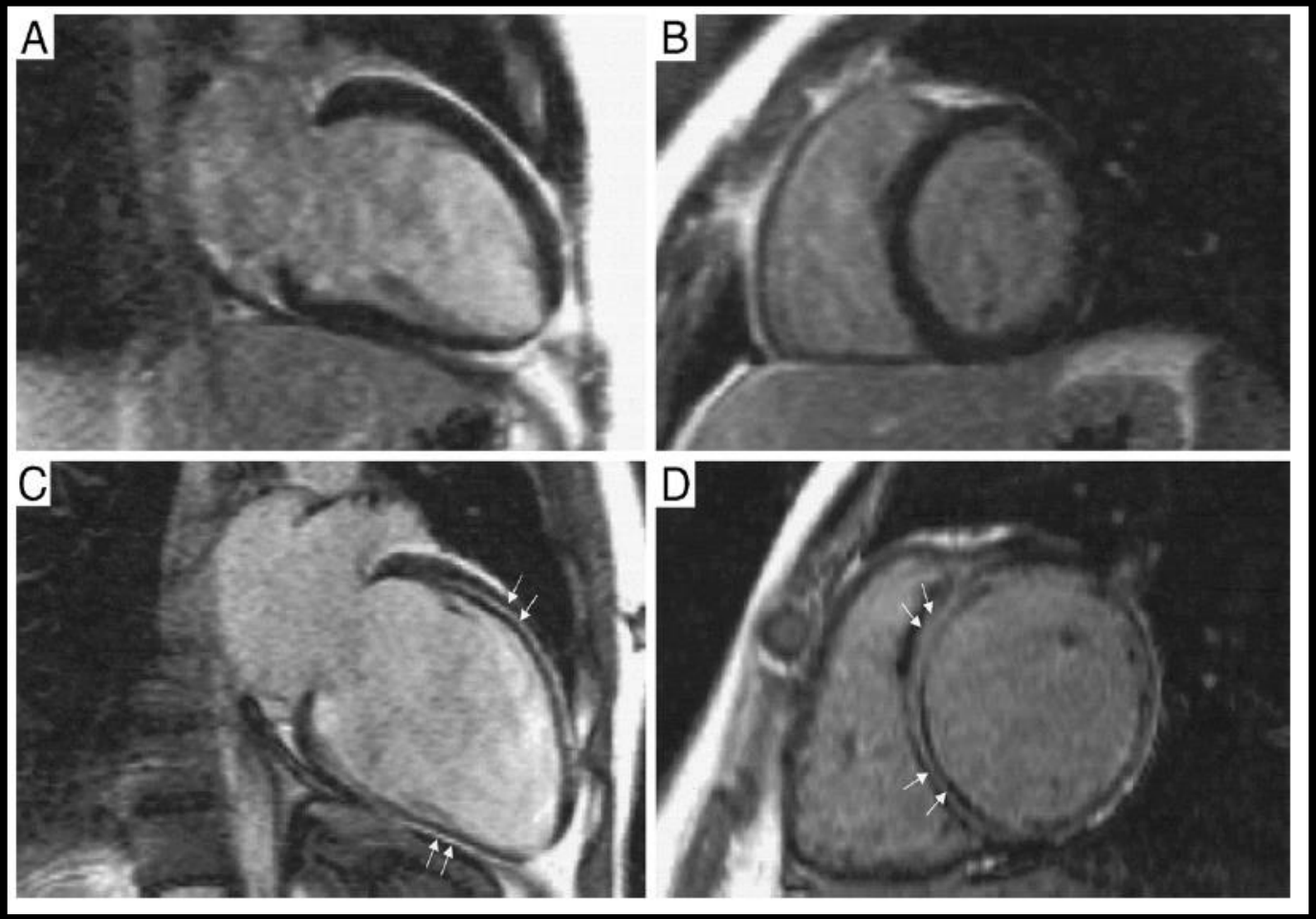
Restrictive



HCM



Dilated Cardiomyopathy

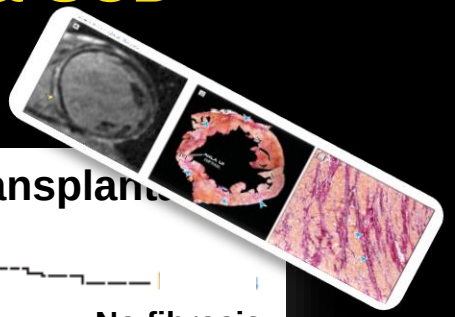


Mid wall enhancement present in 30-35% of dilated cardiomyopathy

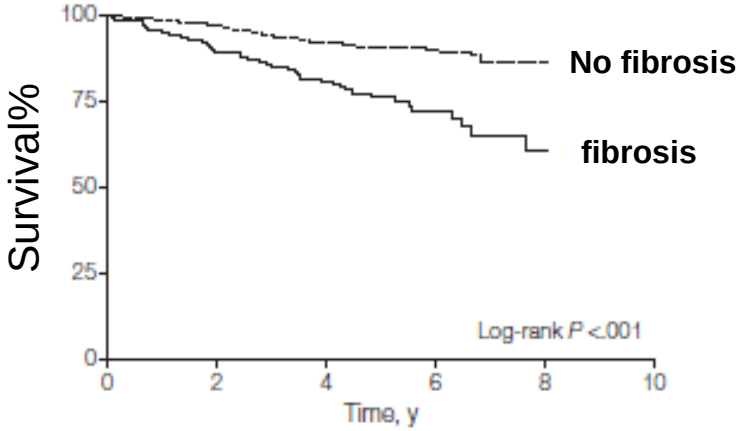
Multivariate analysis showed mid-wall fibrosis is sole predictor of death or hospitalisation

Assomull et al. JACC 2006

Association of Fibrosis with Mortality & SCD
- in Non-ischemic DCM -

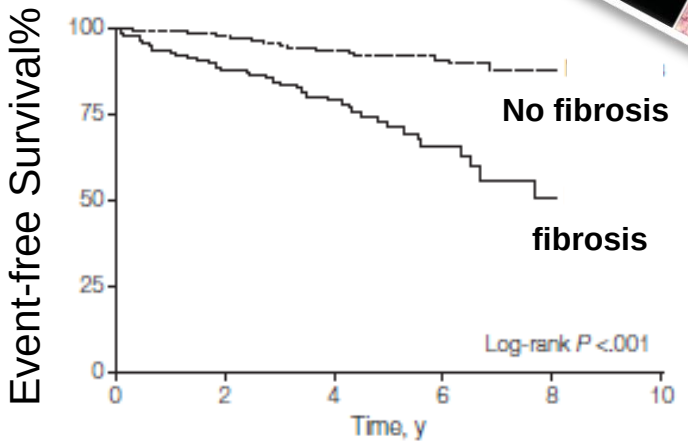


All-cause mortality



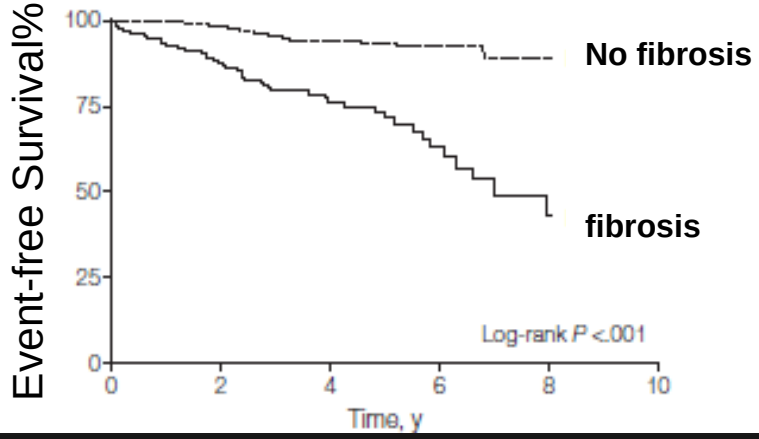
No. at risk					
No fibrosis	330	318	260	136	51
Fibrosis	142	122	99	39	13

CV mortality or transplant

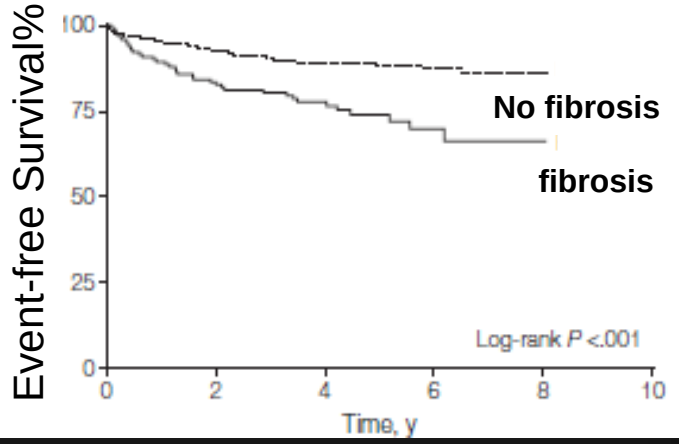


No. at risk					
No fibrosis	330	316	184	93	26
Fibrosis	142	120	79	28	10

Sudden cardiac death or aborted SCD



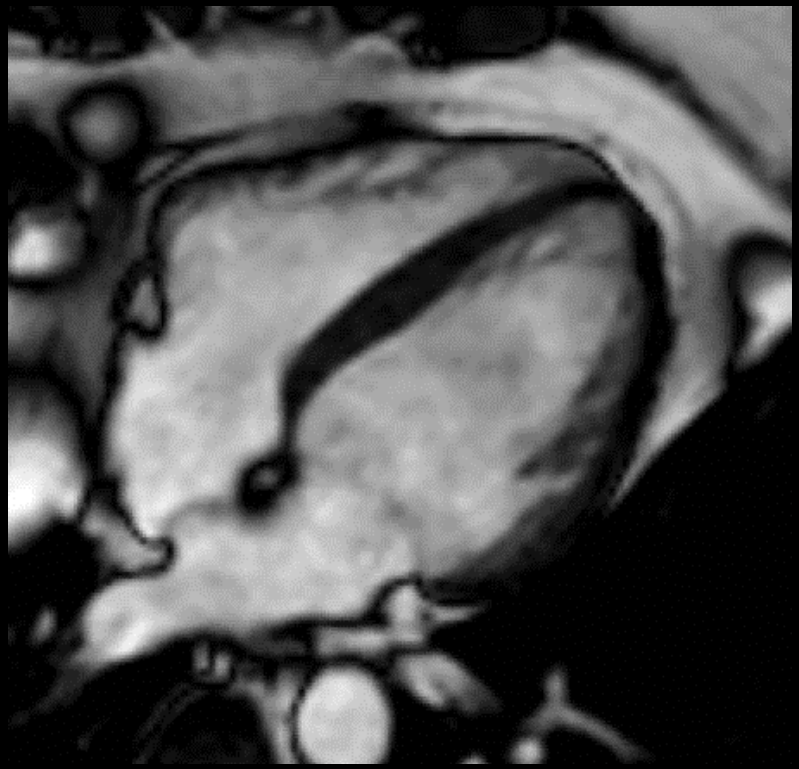
Heart failure, hospitalisation, HTX



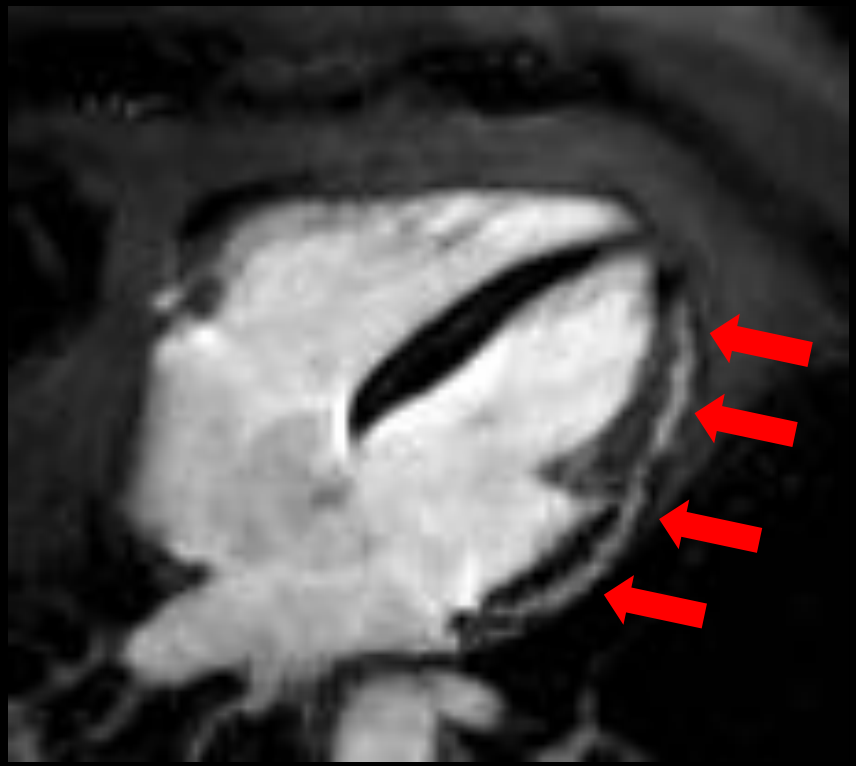
472 pat; FU 5.3 years; Mean EF <40%

Gulati et al. JAMA 2013

Neuromuscular disorders
- limb-girdle muscular dystrophy -

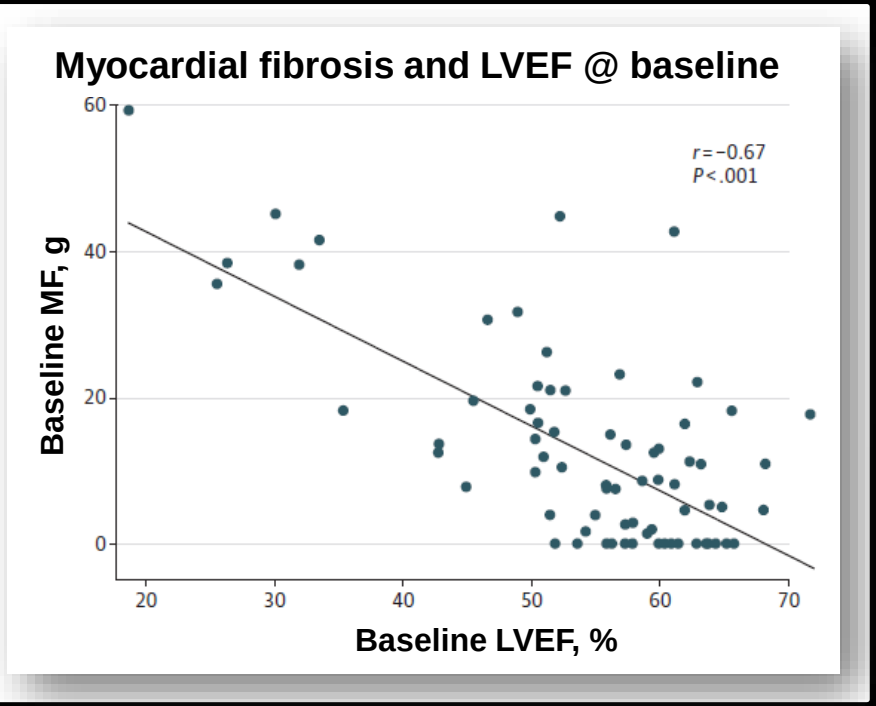


Cine

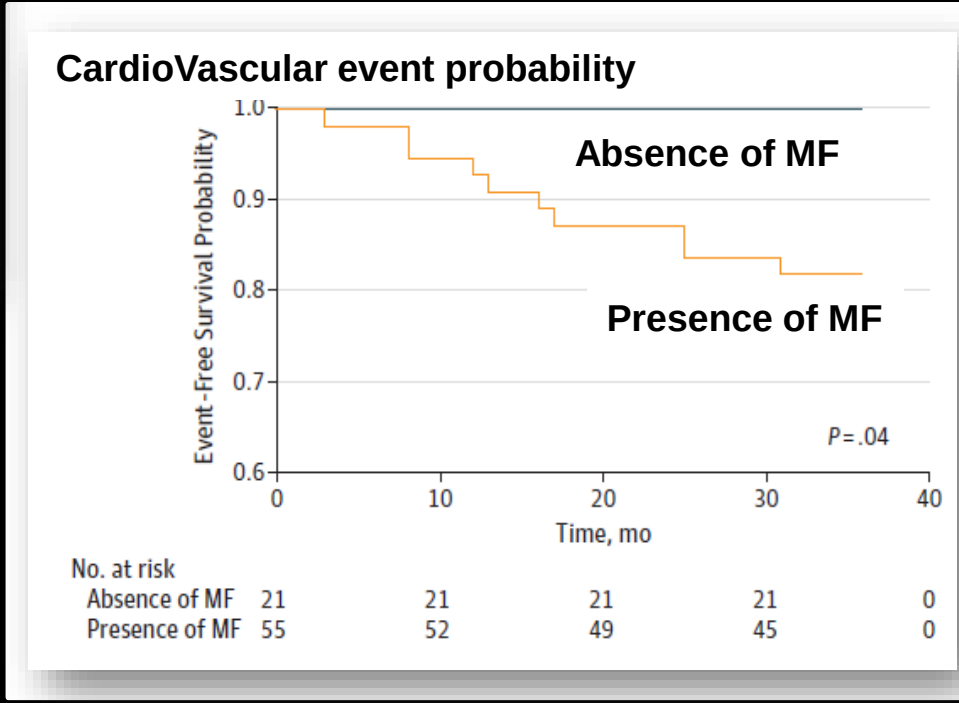


Late enhancement

Neuromuscular disorders
- Duchenne and Becker Dystrophy -



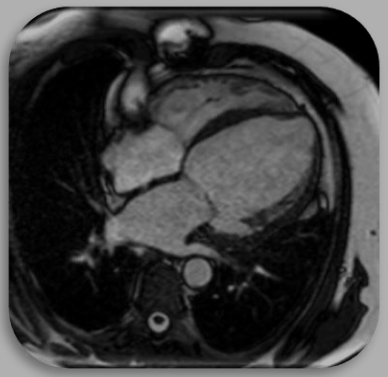
MF: Myocardial fibrosis



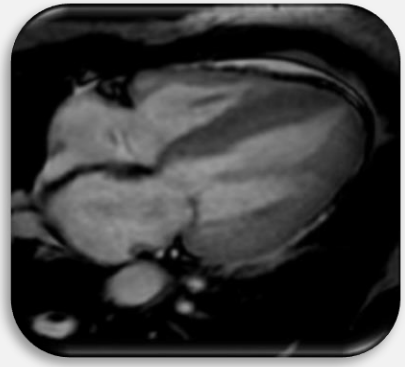
Silva et al. JAMA Cardiol 2017

Non-Ischemic Cardiomyopathies

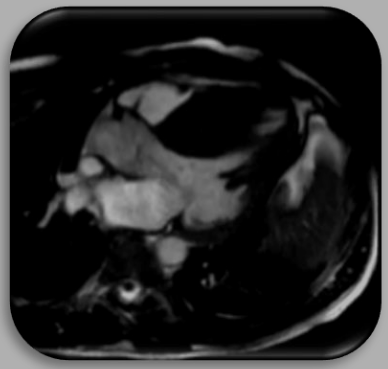
Dilated



Restrictive

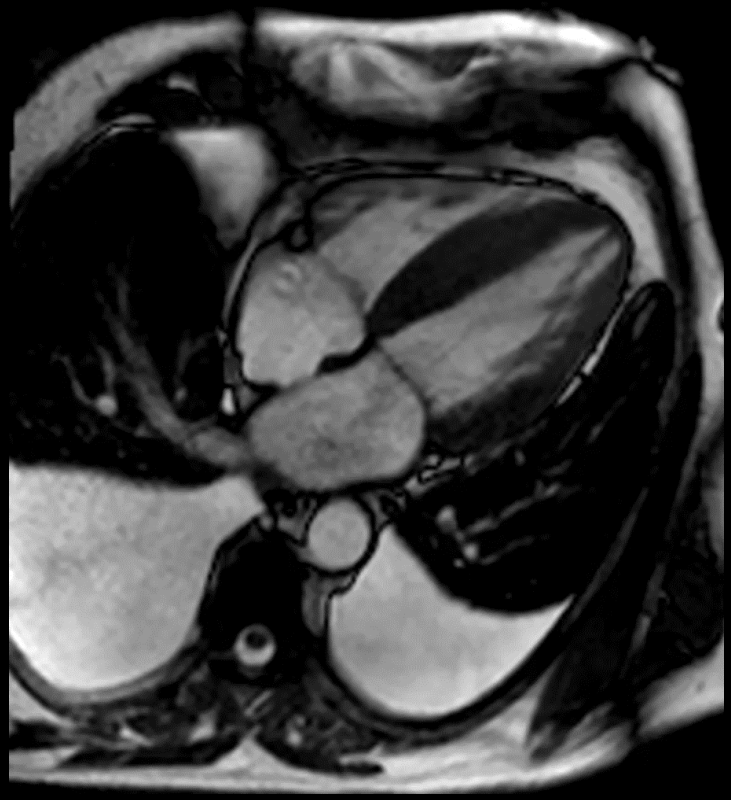


HCM

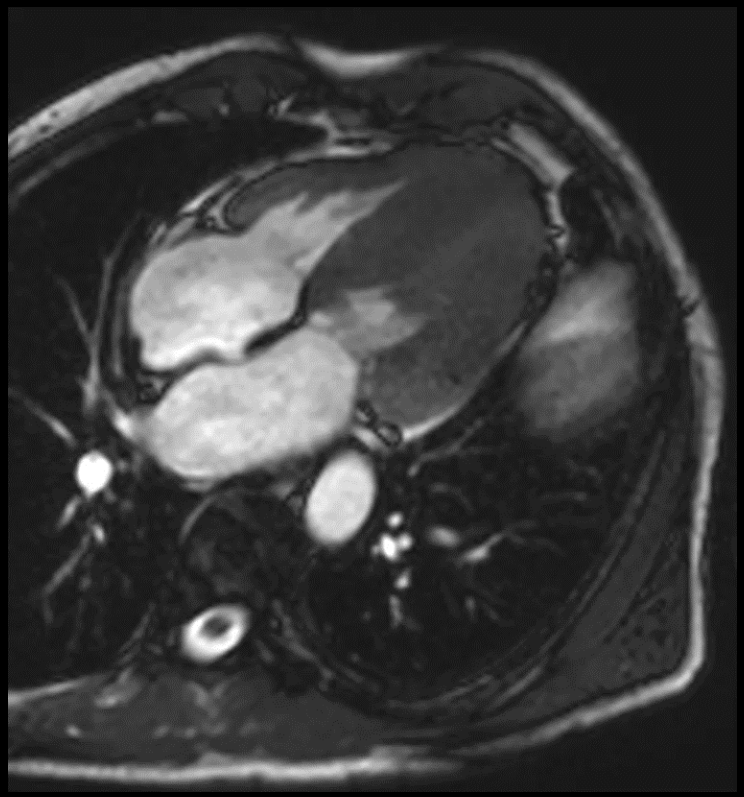


Cardiomyopathy
- Diagnosis & Etiology-

59 yo, male, dyspnea (NYHA III)

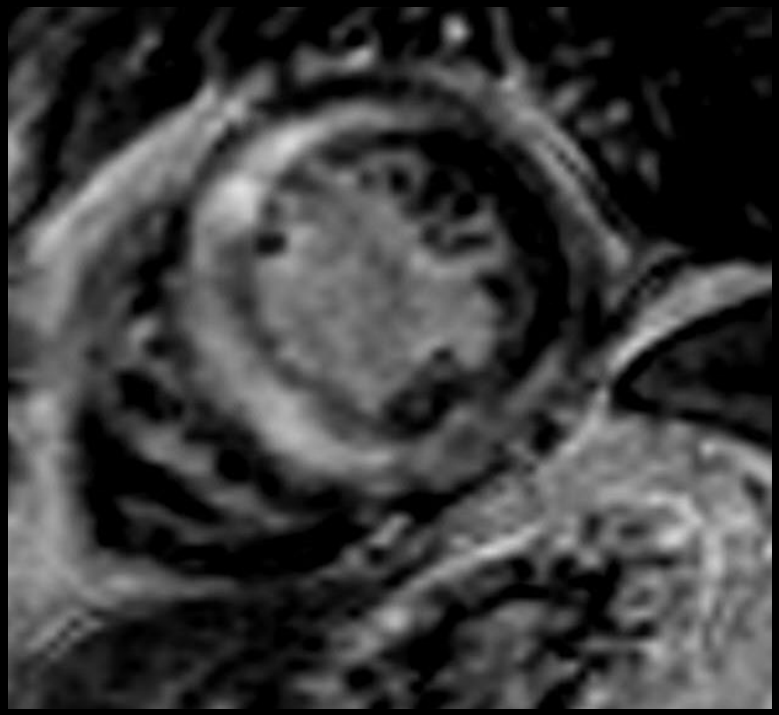


59 yo, male, dyspnea (NYHA II)

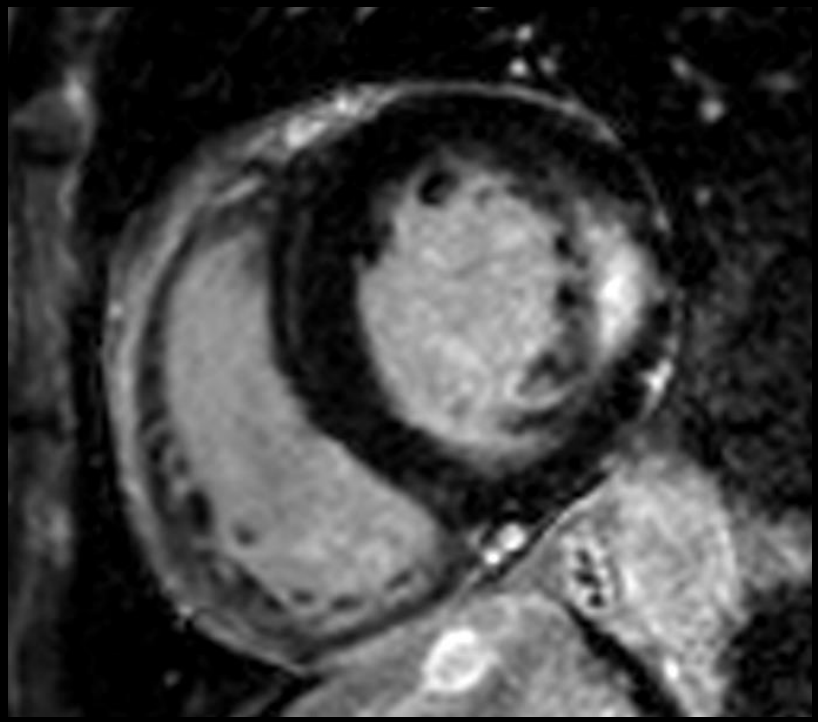


Cardiomyopathy
- Diagnosis & Etiology-
Late Gadolinium Enhancement

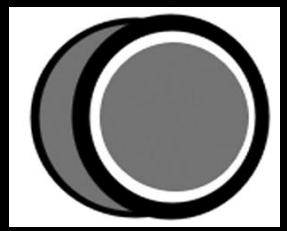
59 yo, male, dyspnea (NYHA III)



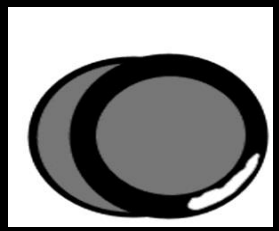
59 yo, male, dyspnea (NYHA II)



Amyloidosis



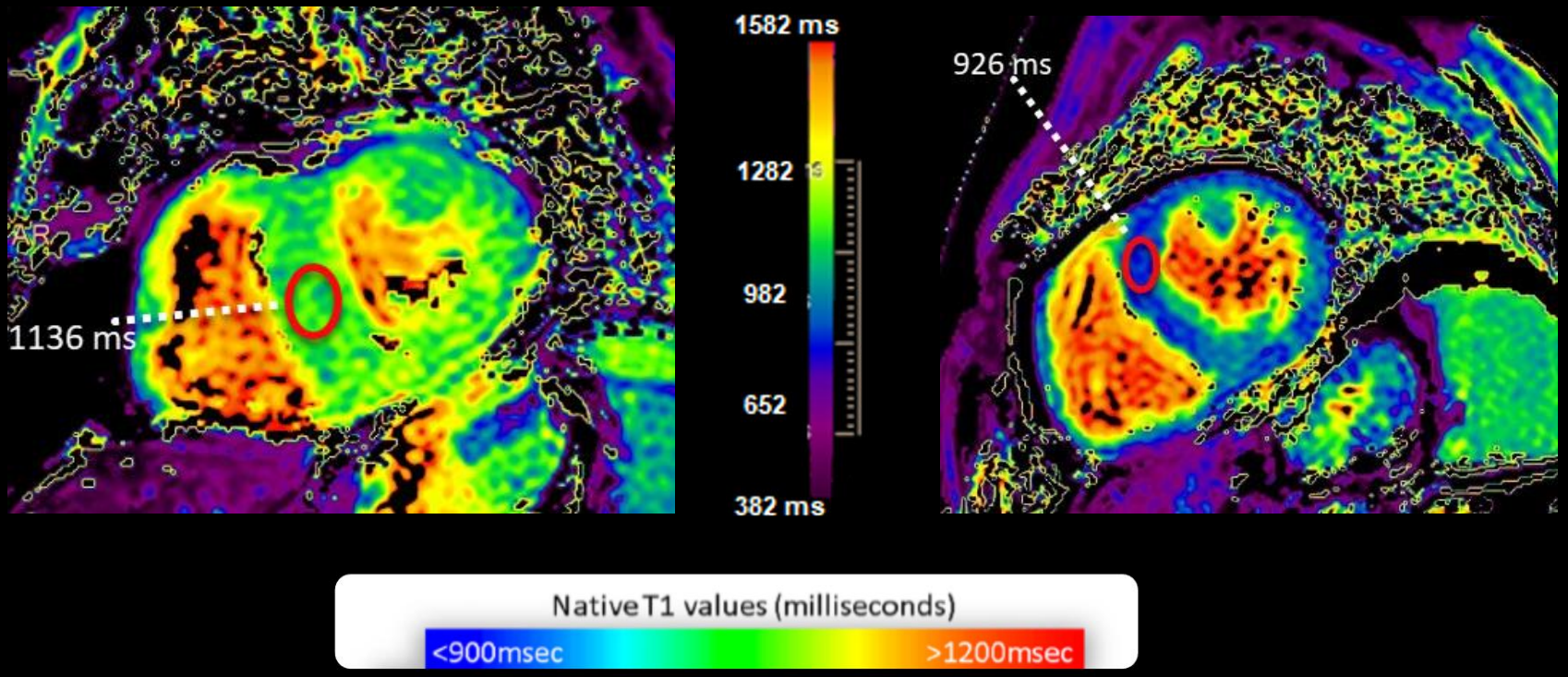
Morbus Fabry



Cardiomyopathy
- Diagnosis & Etiology-
T1- Mapping

59 yo, male, dyspnea (NYHA III)

59 yo, male, dyspnea (NYHA II)

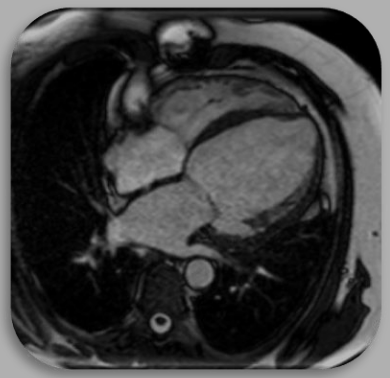


Amyloidosis

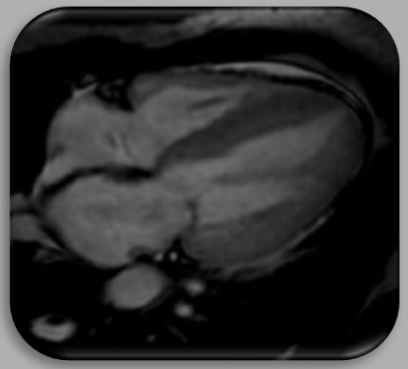
Morbus Fabry

Non-Ischemic Cardiomyopathies

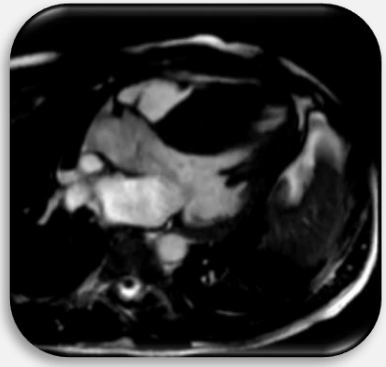
Dilated



Restrictive

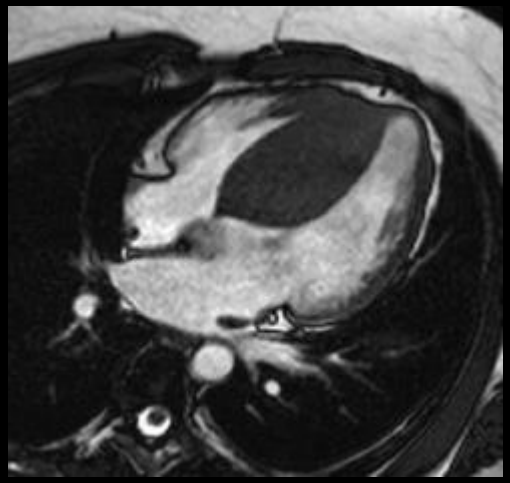


HCM

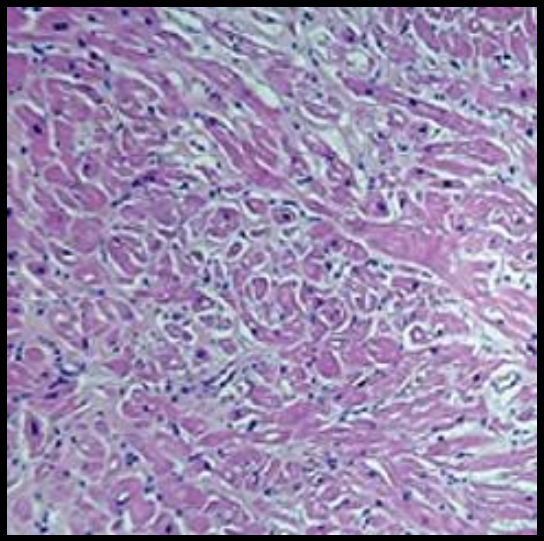


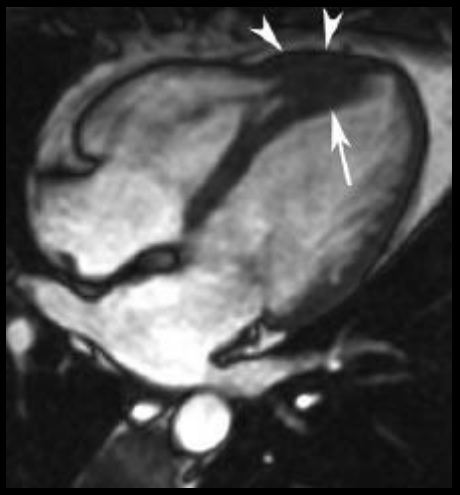
Hypertrophic Cardiomyopathy

- Phenotypes -

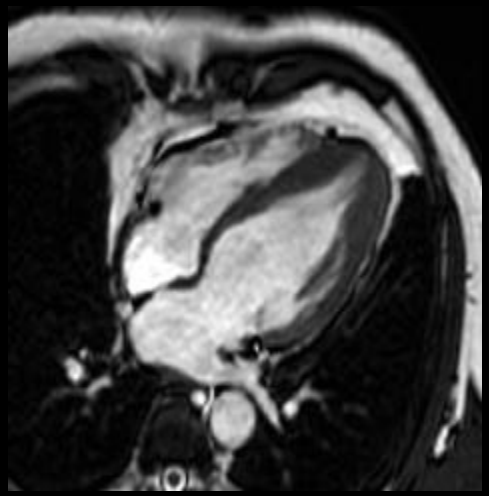


Septal

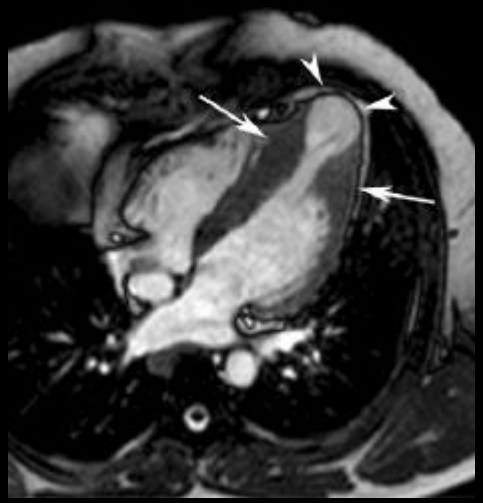




Focal



Apical

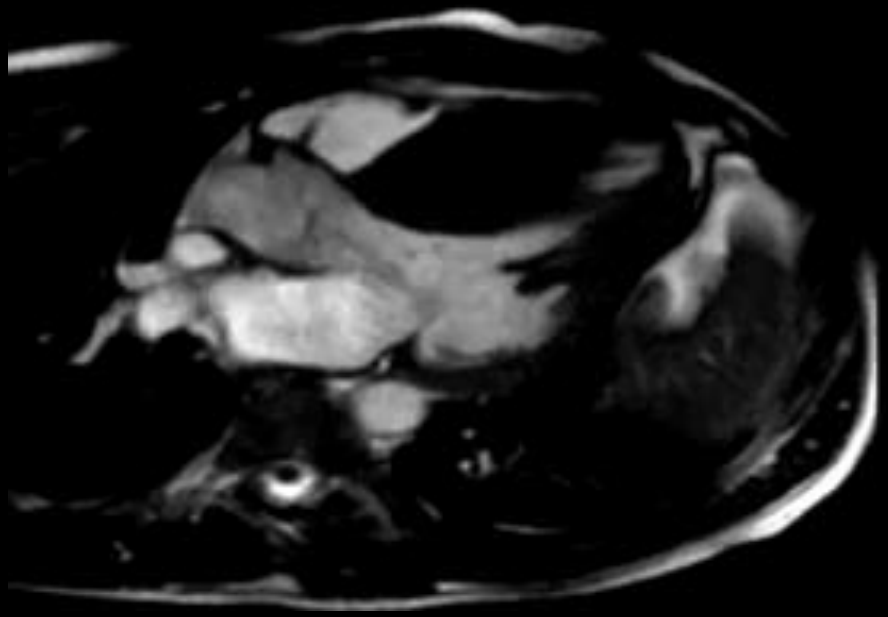


Midventricular

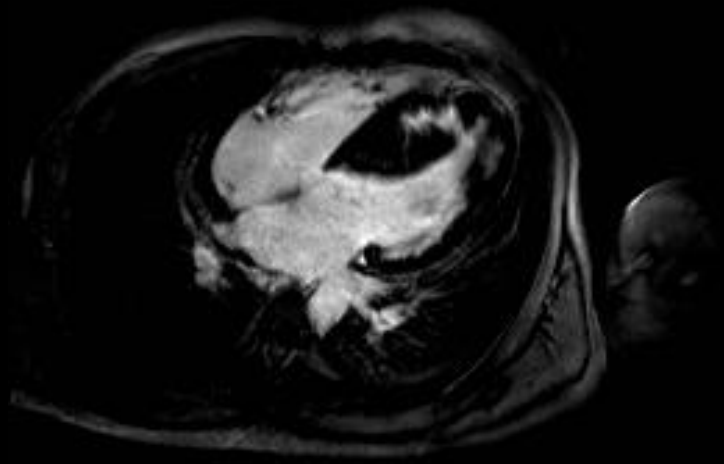
Bogeart et al. Radiology 2014

Hypertrophic (obstructive) Cardiomyopathy

Cine



Late Enhancement

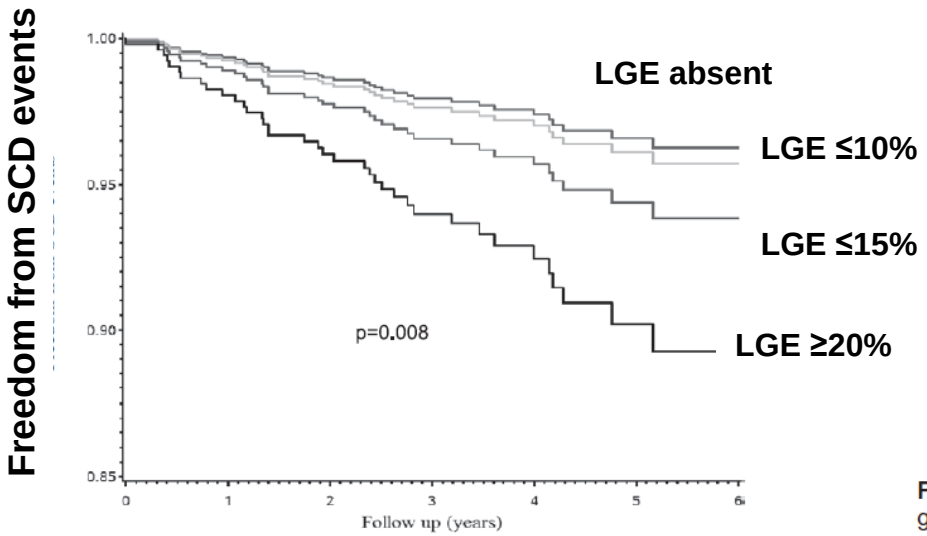


Male 25 yo

Max. septal Wall 32 mm

Total amount of LV- Fibrosis 7%

Prognostic value of CMR in HCM



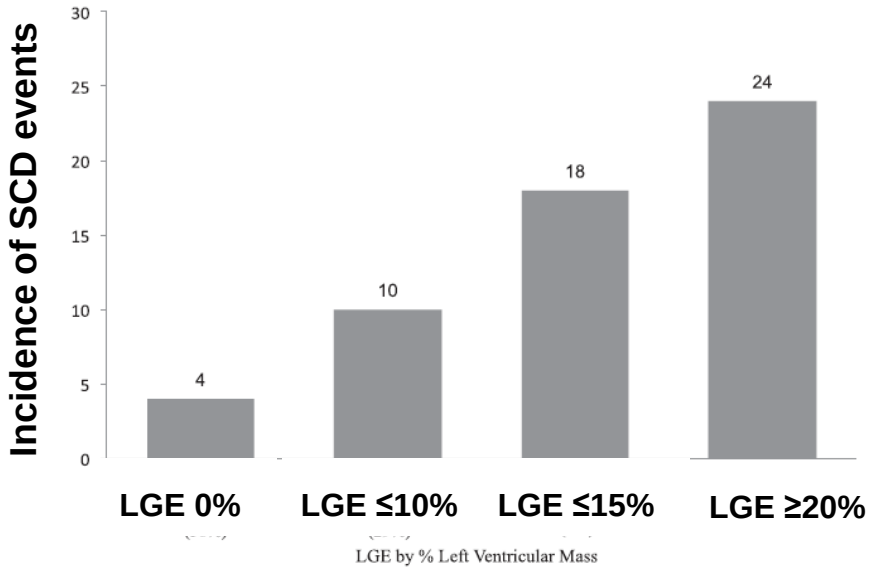
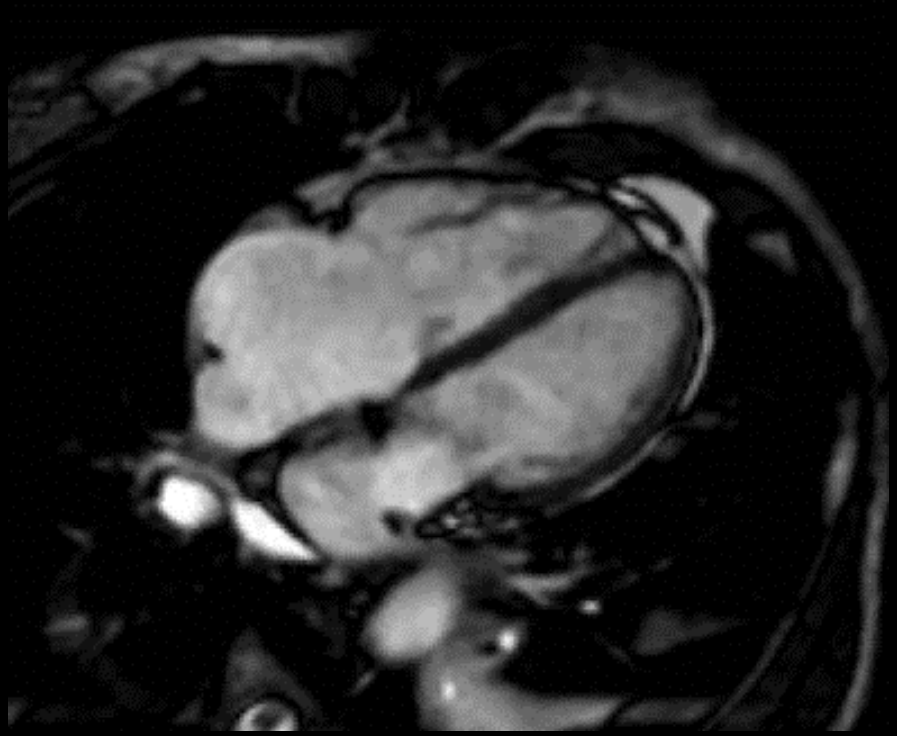


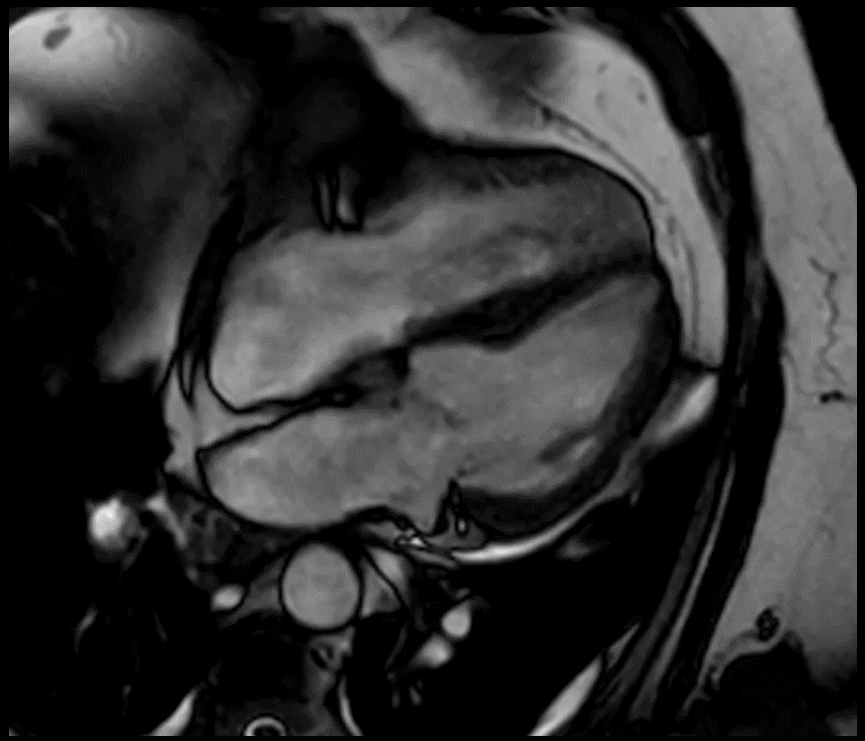
Figure 2. Relation between extent of late gadolinium enhancement (LGE) and sudden cardiac death (SCD) events in 1293 patients with hypertrophic cardiomyopathy. **A**, Hazard plot based on multivariable Cox regression analysis (P=0.008). **B**, Incidence of SCD events increased progressively and in direct relation to the extent of LGE (P<0.001).

Chan et al, Circulation 2014

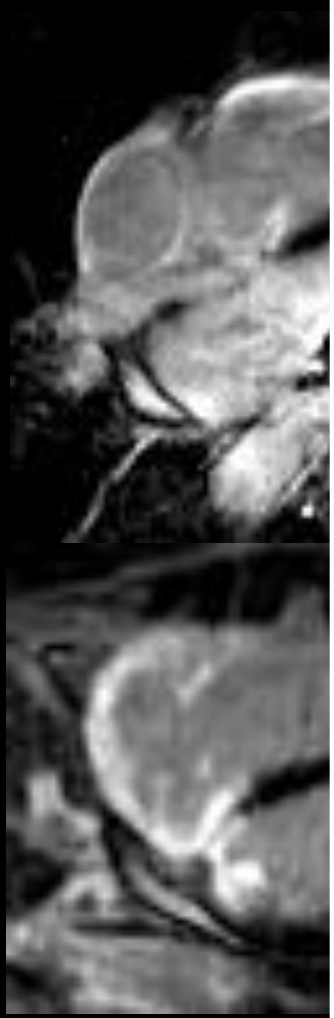
Cardiomyopathy
- Diagnosis & Etiology-



60 yo female
LVEF 50%
RVEF 25%
NSVTs

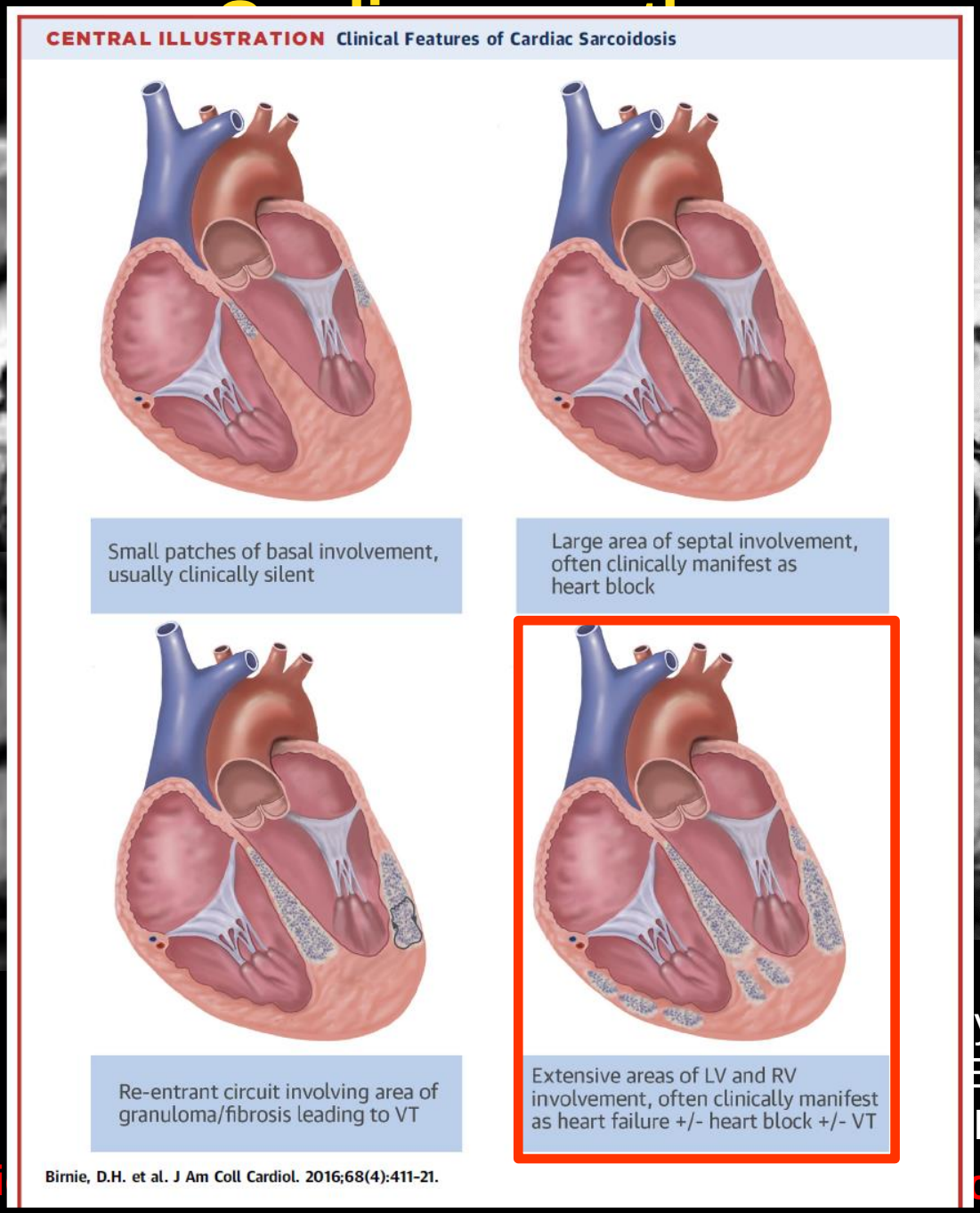


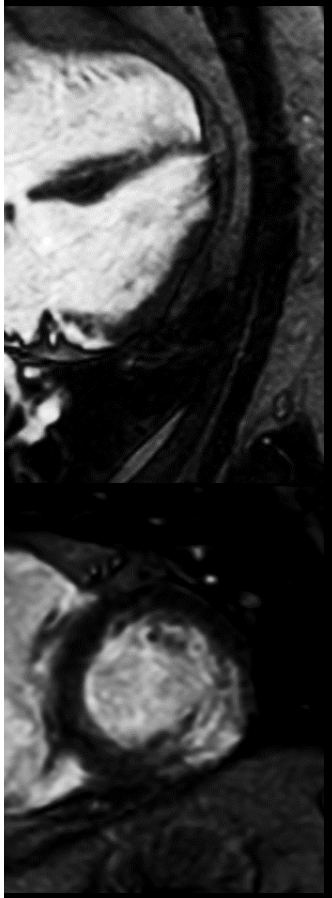
65 yo male
LVEF 48%
RVEF 31%
VTs



60 yo female
LVEF 50%
RVEF 25%

RV-Infarcti

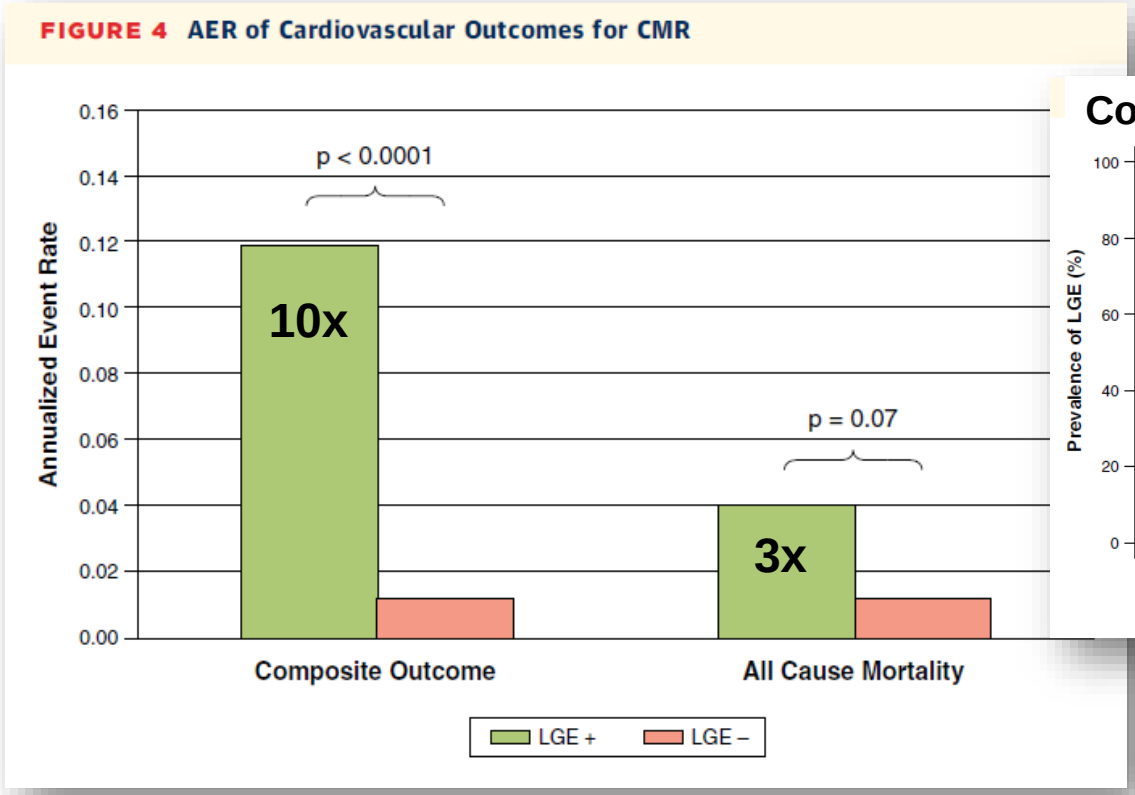


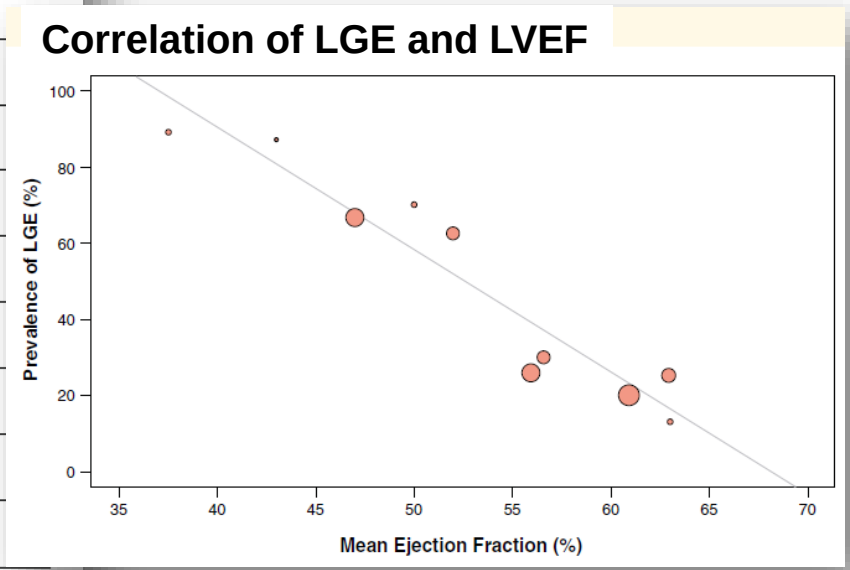


yo male
EF 48%
EF 31%

oidosis

Prognosis - MRI
- Sarcoidosis -





Composite outcome: Mortality and arrhythmogenic events
AER: Annualized Event Rate
Pooled Data: 10 studies – 760 patients

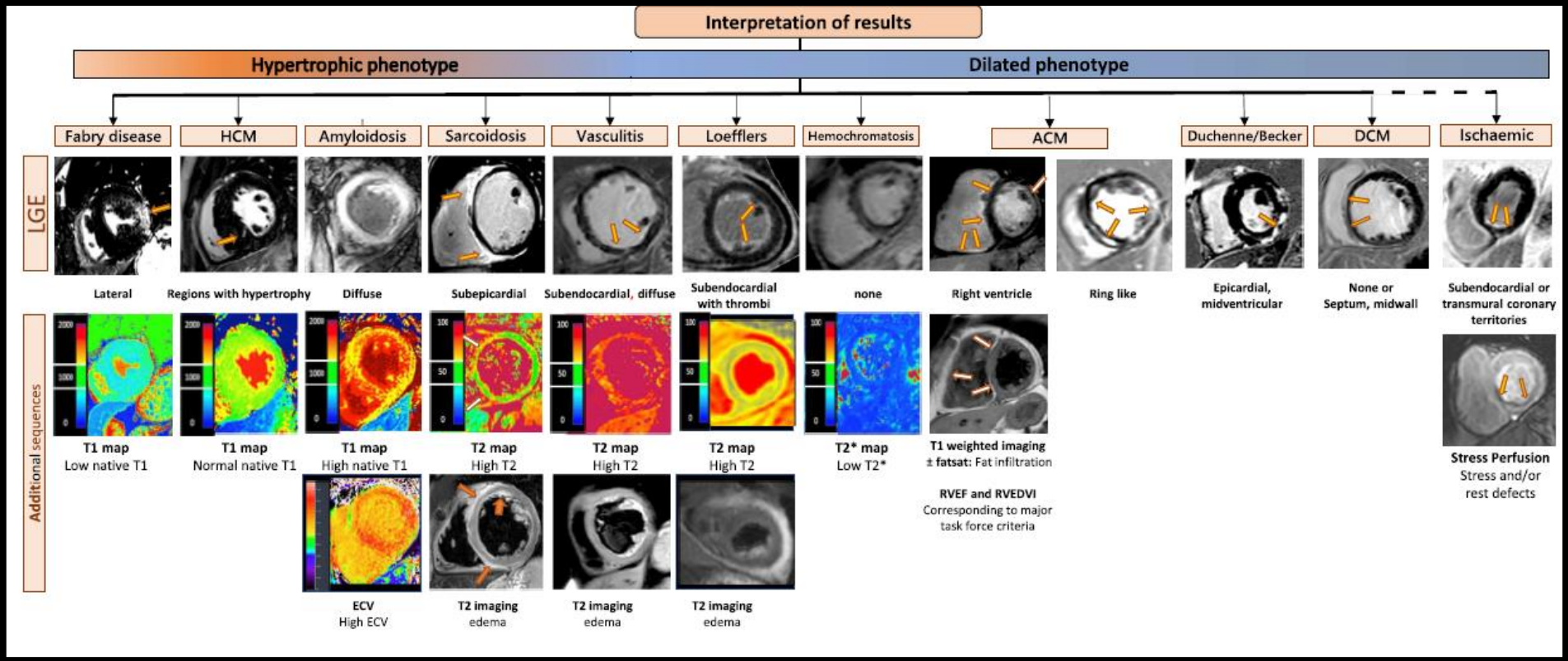
Colemann et al. JACCimg 2017

Interpretation of Results

ESC European Society of Cardiology
European Heart Journal - Cardiovascular Imaging (2022) 23, 587–589
https://doi.org/10.1093/ehjci/jeac051
HOW TO

How to evaluate cardiomyopathies by cardiovascular magnetic resonance parametric mapping and late gadolinium enhancement

Nassiba Menghoum[1], Jacqueline L. Vos[2], Anne-Catherine Pouleur[1], Robin Nijveldt[2], and Bernhard L. Gerber [1]*



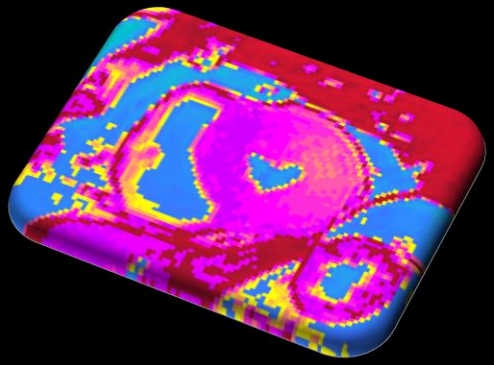
Conclusion

Cardiac MRI in Cardiomyopathies:

- **Has a ESC Class I Indication**
- **Differentiates ischemic fro**
- **p** **in the establishment of diagnosis and etiology**
  - **Characterization of Cardiomyopathies**
- **Provides strong prognostic information**

AI might enable deeper phenotyping

Acknowledgements



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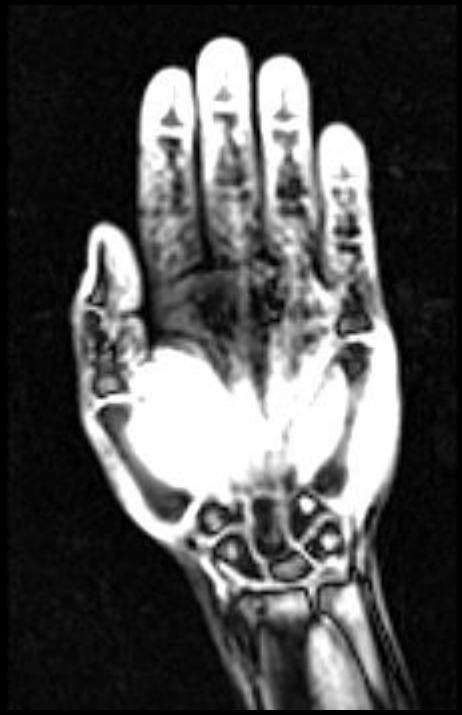

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