EACVI CMR Exam Prep Course

HQ European Society of Cardiology

Nice, 29-30 Sept 2022

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European Association of
Cardiovascular Imaging

HCM and ARVC

Chiara Bucciarelli-Ducci, MD, PhD, FESC, FRCP

29 September 2022

ESC
European Society
of Cardiology

COI

- CEO (part-time) Society for Cardiovascular Magnetic Resonance
- Speaker’s fees Circle Cardiovascular Imaging, Bayer, Siemens Healthineers.

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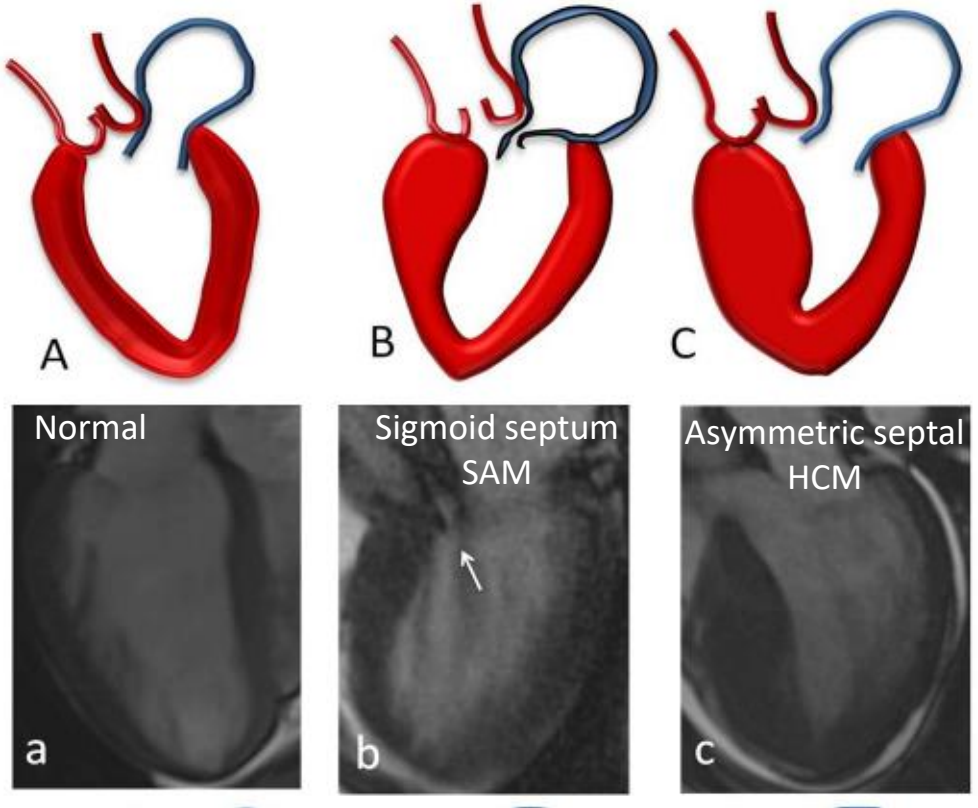
1/ Various patterns of hypertrophy



Noureldin et al JCMR 2012

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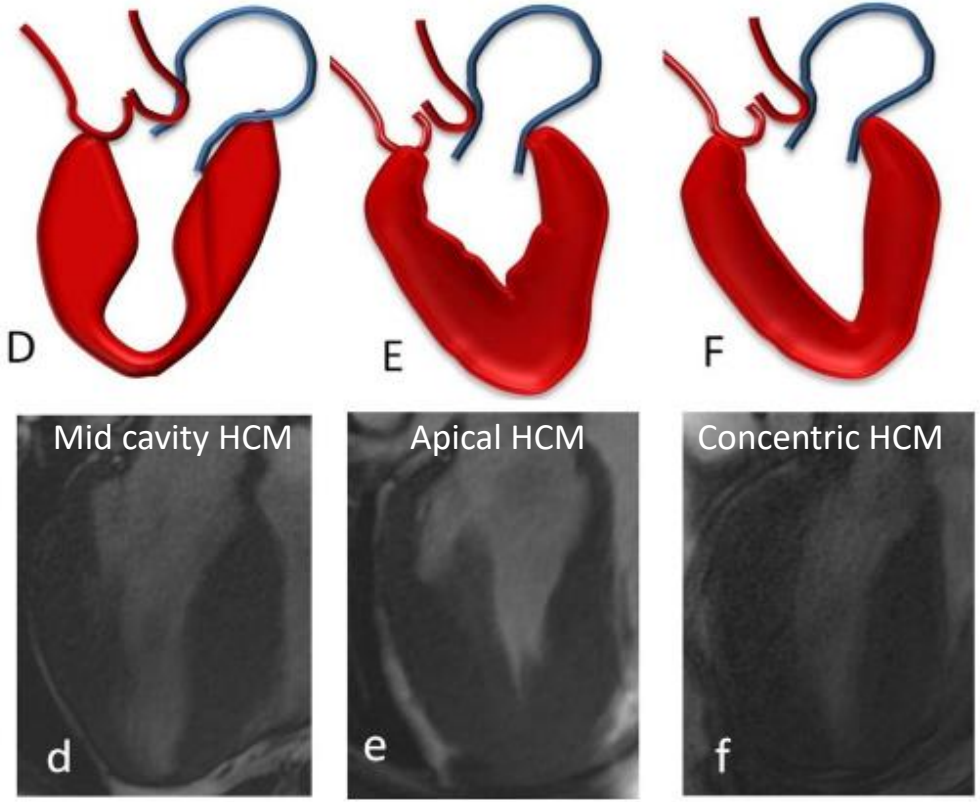

Noureldin et al JCMR 2012

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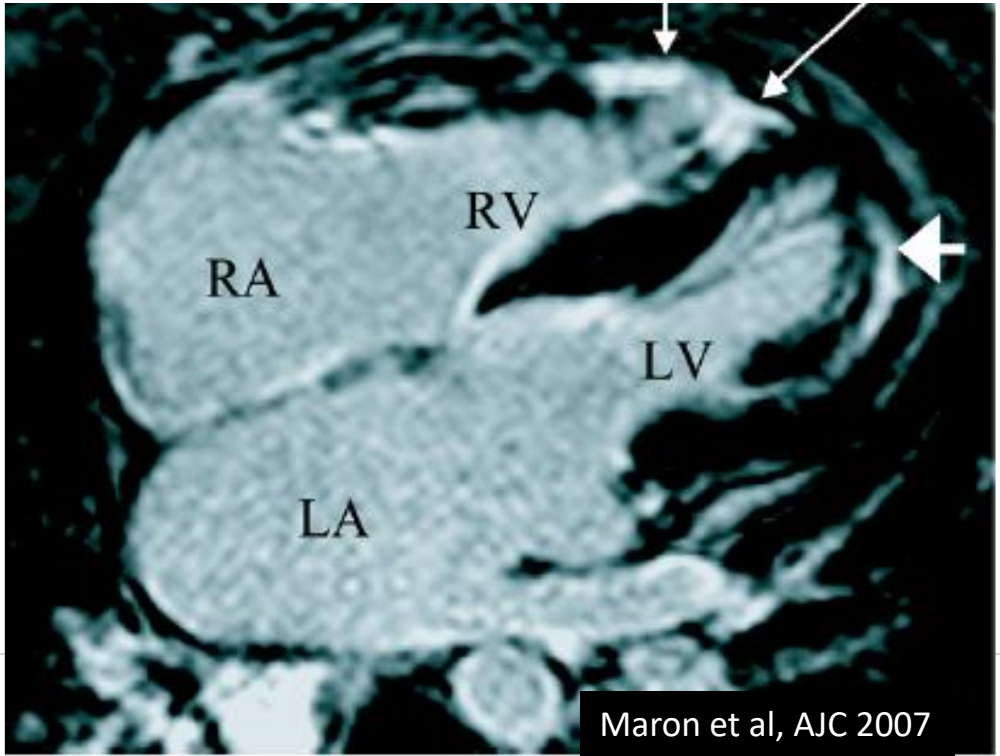
2/RV involvement

Incidence of RV involvement:
not well documented

Incidence of RV involvement: 33%
(RV thickness > 8mm)
Maron et al AJC 2007

Right Ventricular Involvement in Hypertrophic Cardiomyopathy

Martin S. Maron, MD[a,*], Thomas H. Hauser, MD, MMSc, MPH[b], Ethan Dubrow, BA[a],
Taylor A. Horst, BA[a], Kraig V. Kissinger, RT[b], James E. Udelson, MD[a], and Warren J. Manning, MD[b,c]



Maron et al, AJC 2007

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3/ Late gadolinium enhancement (LGE) in HCM

- Frequency
- Typical spatial distribution
- Various morphologies

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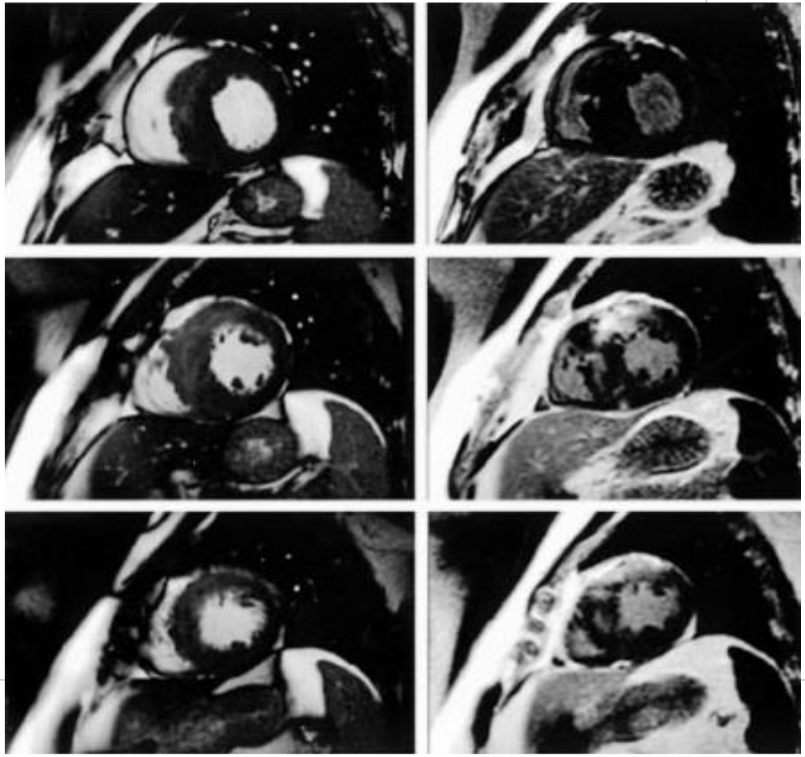
3/ Late gadolinium enhancement (LGE) in HCM

- **Frequency**
- **Typical spatial distribution**
- **Various morphologies**

<u>LGE present in 2/3 cases</u>
BUT depends on Unit volumes/HCM clinics



Moon et al, JACC 2003
Petersen et al, Circulation 2007

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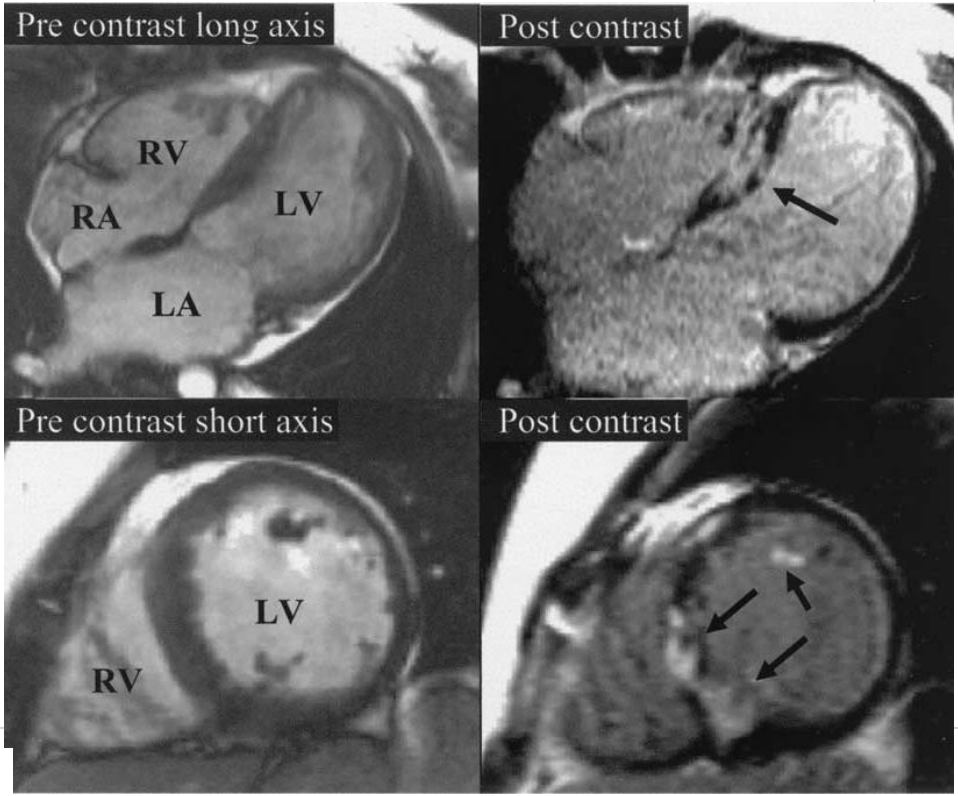
- Frequency
- **Typical spatial distribution**
- **Various morphologies**



Moon et al, JACC 2003

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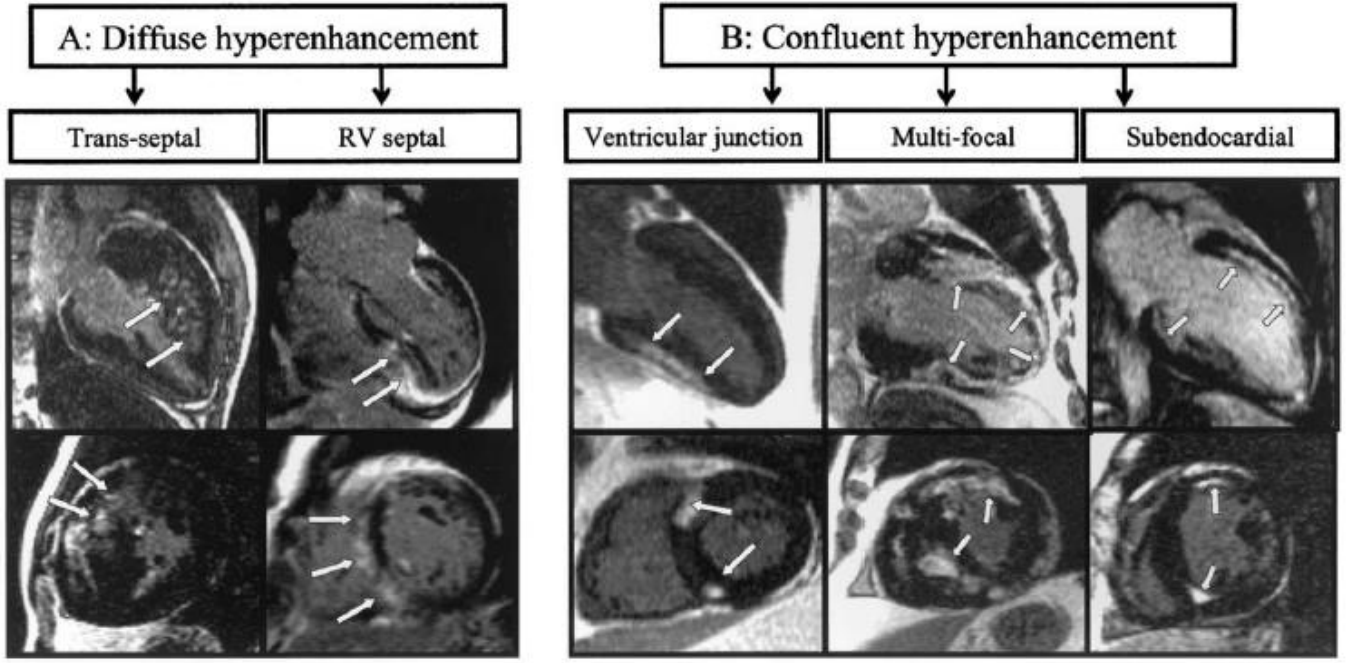


Moon et al, JACC 2003

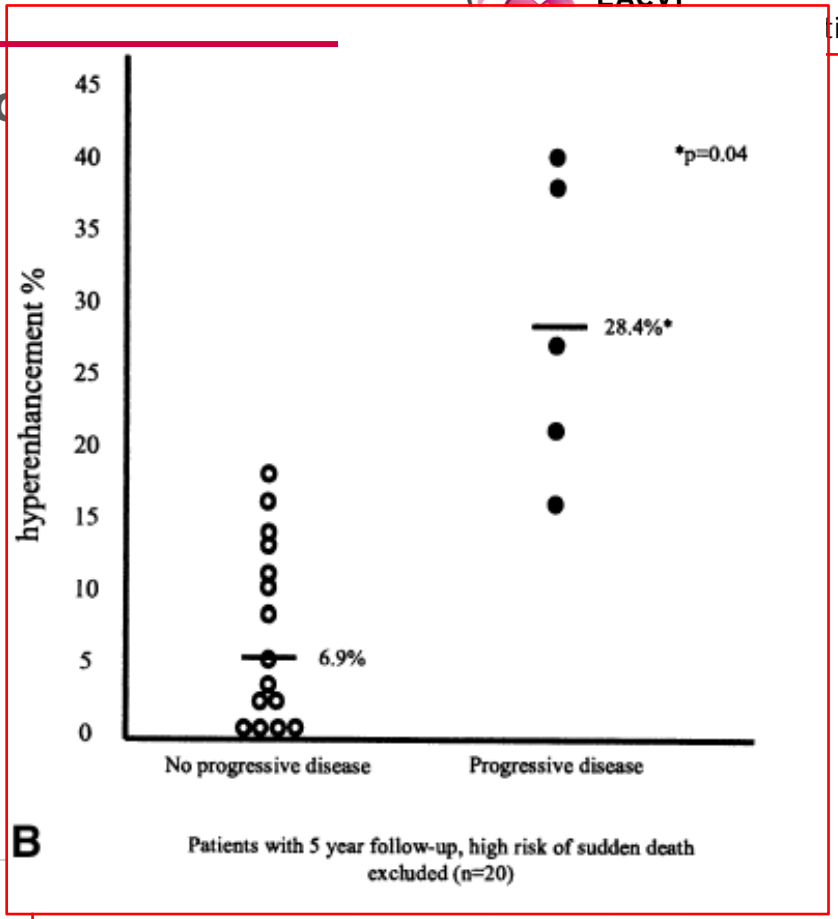
HCM, LGE and Risk Factors

3/ Late gadolinium enhancement (LGE) in HC

- **Frequency**
- **Typical spatial distribution**
- **Various morphologies**

Greater LGE extent in patients with progressive disease (28.5% vs. 8.7%, p 0.001)

Greater LGE extent in patients with two or more risk factors for sudden death (15.7% vs. 8.6%, p 0.02)



Moon et al, JACC 2003

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3/ Late gadolinium enhancement (LGE) in HCM

- Frequency
- Typical spatial distribution
- Various morphologies

Table 2. Associations Between Pattern of Hyperenhancement and Clinical Phenotype

Type of Hyperenhancement	n (%)	Clinical Phenotype
Trans-septal	4 (7%)	Young, gross asymmetric LVH; extensive diffuse hyperenhancement; high risk of sudden death
RV septal	4 (7%)	Extensive RV surface of septal hyperenhancement; strong family history of sudden death
Ventricular junction	12 (23%)	Moderate symmetrical LVH; limited hyperenhancement at RV insertion points; lower risk of sudden death
Multi-focal	9 (17%)	Large focal areas of hyperen-hancement; LBBB if basal septum; associated with progressive disease
Subendocardial	2 (4%)	Like infarcts, but not IHD, in these patients
Other	11 (21%)	Other patterns or trivial hyperenhancement
None	11 (21%)	Typically young and at low risk

IHD = ischemic heart disease; LBBB = left bundle branch block; LVH = left ventricular hypertrophy; RV = right ventricular.

Moon et al, JACC 2003

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3/ Late gadolinium enhancement (LGE) in HCM

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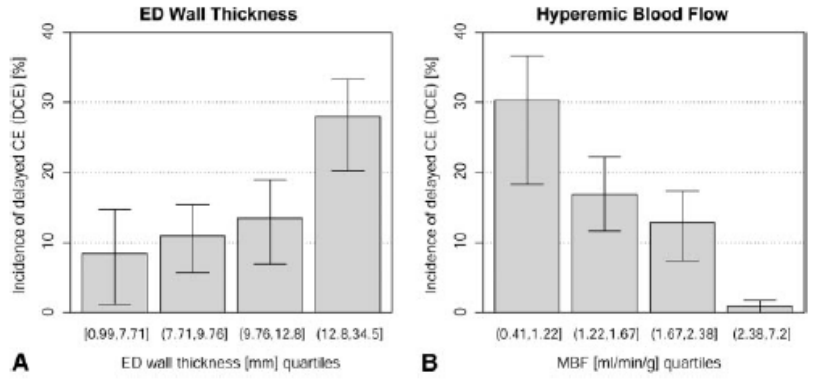
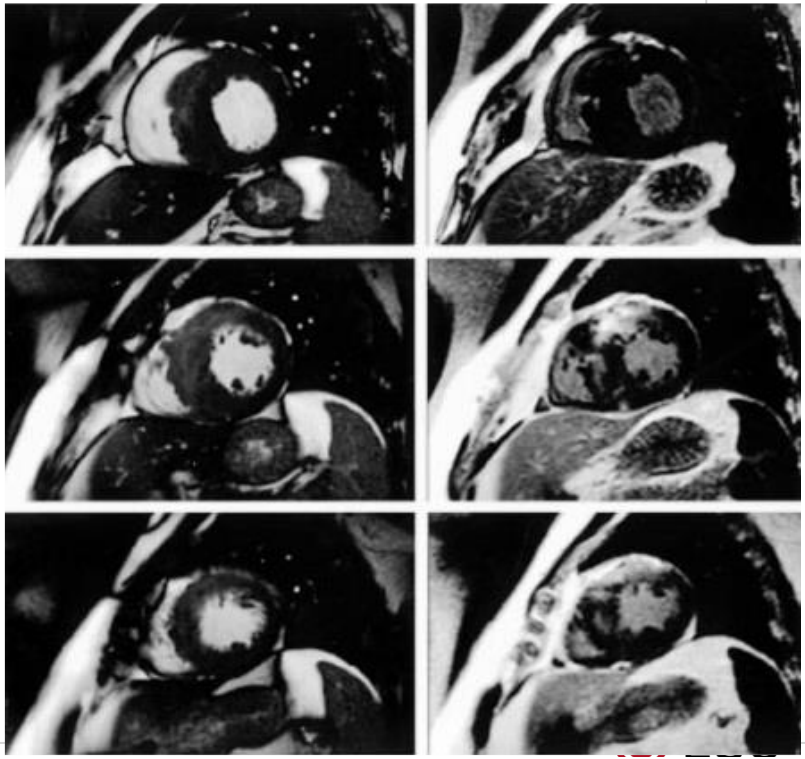
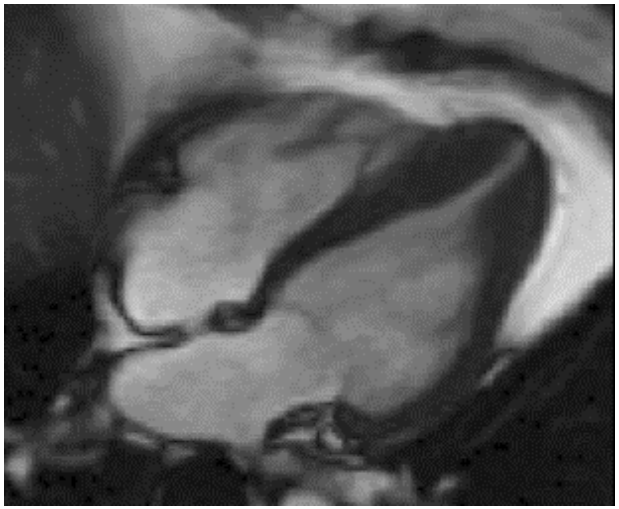


Petersen et al Circulation 2007



Petersen et al Circulation 2007

Apical HCM



Maximum apical thickness by CMR (mm)	Average apical basal ratio by CMR	Echocardiography report
27	1.8	Normal
16	1.4	Trabeculated apex
28	3.2	Akinetic apex
15	1.4	Normal
16	2.0	Normal
20	2.5	Normal
16	2.5	Normal
17	1.7	Normal
24	3.2	Poor views, normal
17	1.9	Normal

Moon et al, Heart 2004

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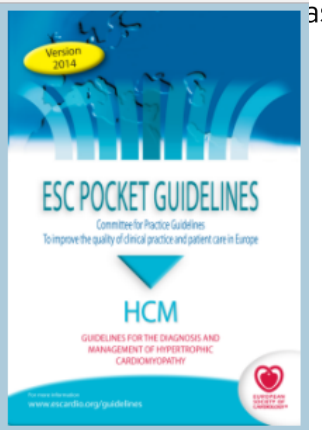
HCM Risk-SCD Calculator

Age	**Years**	*Age at evaluation*
Maximum LV wall thickness	**mm**	*Transthoracic Echocardiographic measurement*
Left atrial size	**mm**	*Left atrial diameter determined by M-Mode or 2D echocardiography in the parasternal long axis plane at time of evaluation*
Max LVOT gradient	**mmHg**	*The maximum LV outflow gradient determined at rest and with Valsalva provocation (irrespective of concurrent medical treatment) using pulsed and continuous wave Doppler from the apical three and five chamber views. Peak outflow tract gradients should be determined using the modified Bernouilli equation: Gradient= $4V^2$, where V is the peak aortic outflow velocity*
Family History of SCD	○ No ○ Yes	*History of sudden cardiac death in 1 or more first degree relatives under 40 years of age or SCD in a first degree relative with confirmed HCM at any age (post or ante-mortem diagnosis).*
Non-sustained VT	○ No ○ Yes	*3 consecutive ventricular beats at a rate of 120 beats per minute and <30s in duration on Holter monitoring (minimum duration 24 hours) at or prior to evaluation.*
Unexplained syncope	○ No ○ Yes	*History of unexplained syncope at or prior to evaluation.*

Risk of SCD at 5 years (%):	
ESC recommendation:	

Reset



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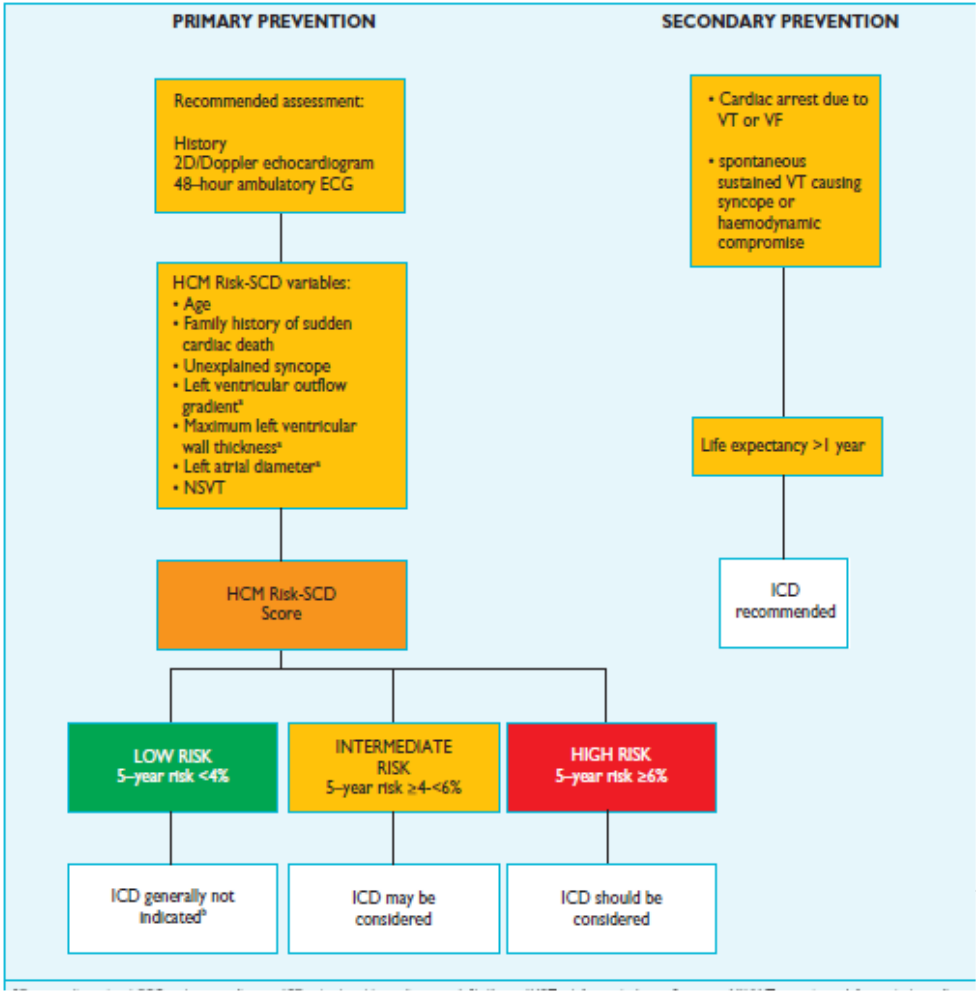
4/ CMR findings and indication for ICD

2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy

http://www.doc2do.com/hcm/webHCM.html



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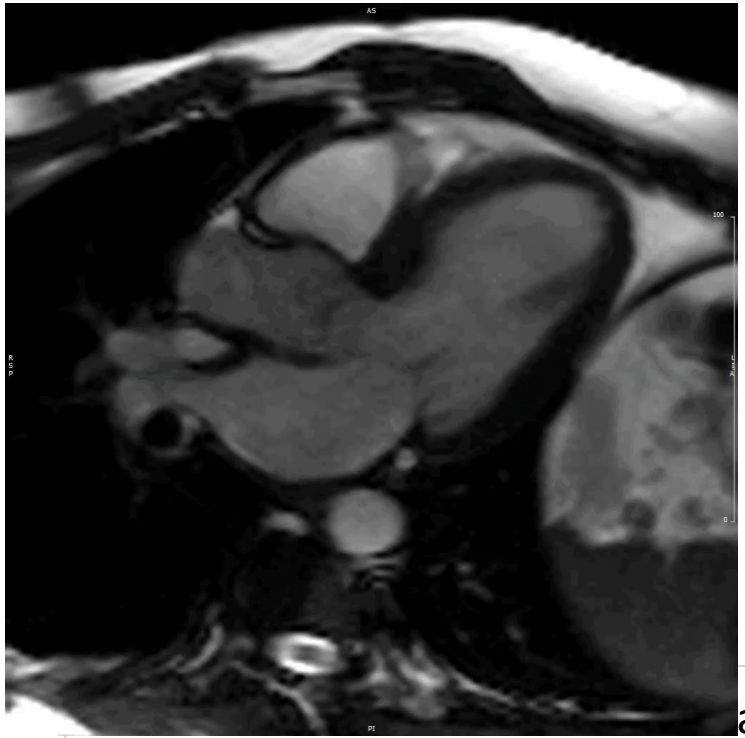
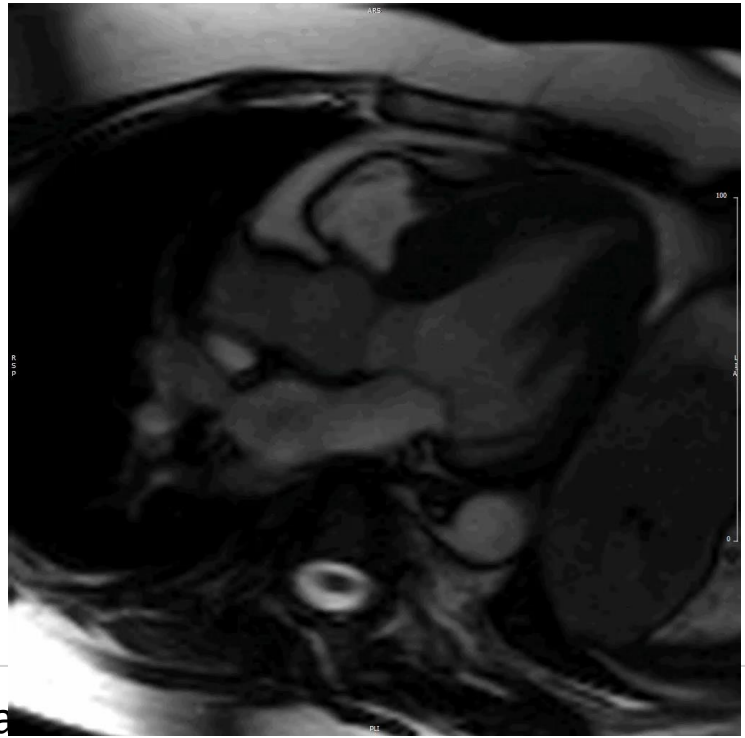
5/Mitral valve in HCM





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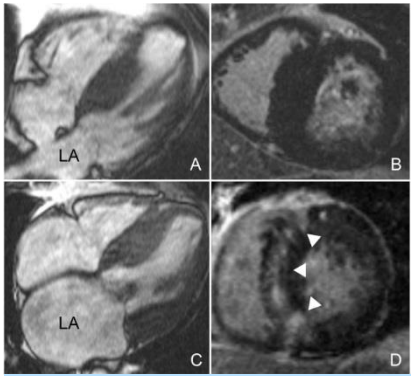
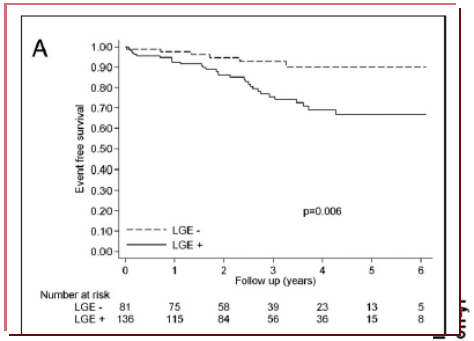
6/CMR findings in HCM and prognosis





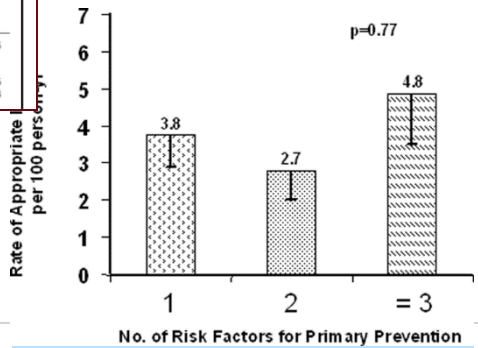
LGE and LA volumes:
Predictors of AF and HF

Hypertrophic Cardiomyopathy

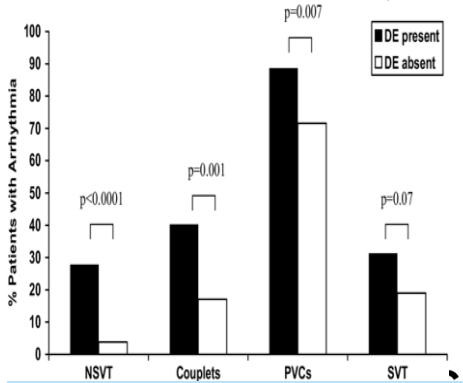
Prognostic Significance of Myocardial Fibrosis in Hypertrophic Cardiomyopathy

Rory O'Hanlon, MD,* Agata Grasso, MD,* Michael Roughton, MSc,¶ James C. Moon, MD,§
Susan Clark, RN,* Ricardo Wage,* Jessica Webb, MD,* Meghana Kulkarni, MD,*
Dana Dawson, MD, PHD,* Leena Sulaibeekh, MD,* Badri Chandrasekaran, MD,*
Chiara Bucciarelli-Ducci, MD,* Ferdinando Pasquale, MD,§ Martin R. Cowie, MD,†
William J. McKenna, MD,‖ Mary N. Sheppard, MD,‡ Perry M. Elliott, MD,‖
Dudley J. Pennell, MD,* Sanjay K. Prasad, MD*
London, United Kingdom

O'Hanlon et al, JACC 2010



ICD Approp Discharge Rate



Presence of LGE:
Risk for ventricular arrhythmias

Differential Diagnosis

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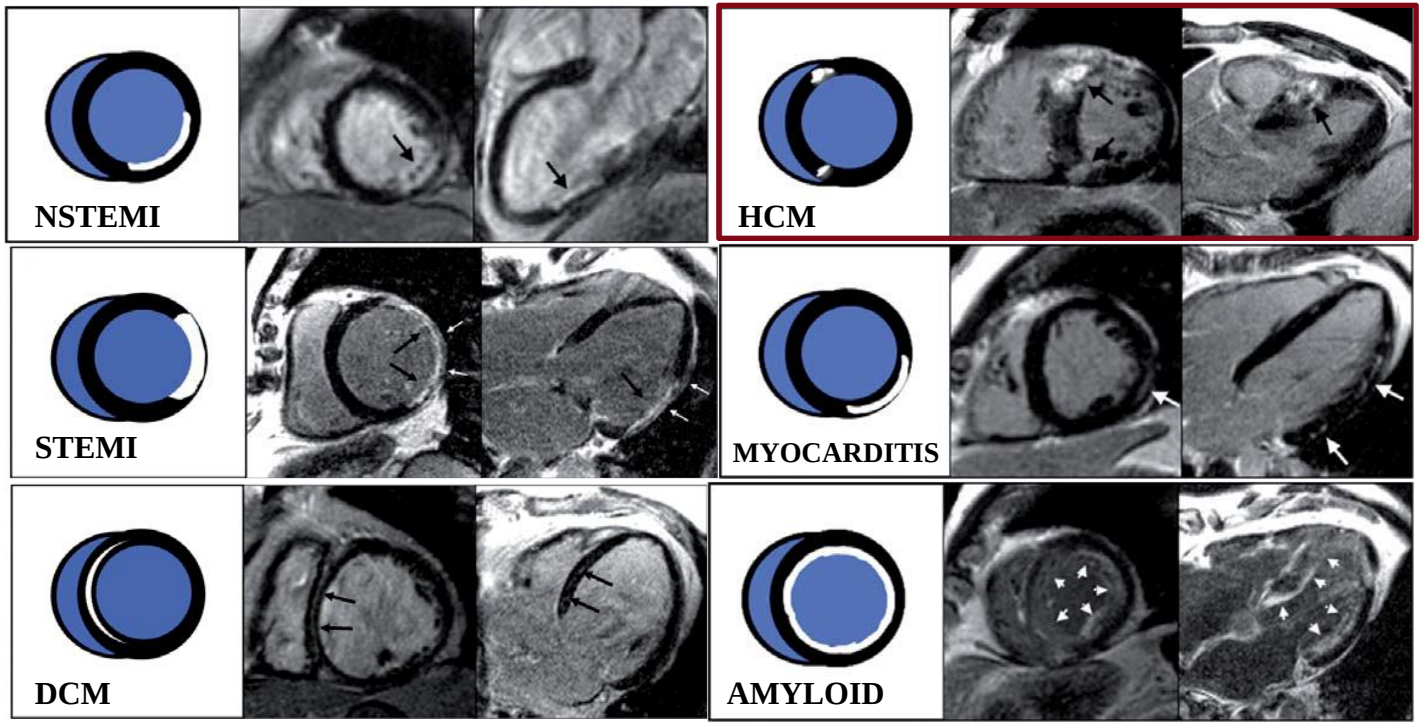

From Bright is Dead to…… Bright is BAD

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Bucciarelli-Ducci C, JACC 2012

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7/Left atrium and HCM

2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy

- Prognostically important
- Anteroposterior left atrial diameter (LA volume indexed for BSA)
- Cause left atrium enlargement is multifactorial
  - SAM and mitral regurgitation
  - Impaired LV filling
  - Concomitant hypertensive heart disease

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8/Relative diagnostic yields for HCM by echo and CMR

Circ Cardiovasc Imaging. 2017 Aug;10(8). pii: e006309. doi: 10.1161/CIRCIMAGING.117.006309.

Discrepant Measurements of Maximal Left Ventricular Wall Thickness Between Cardiac Magnetic Resonance Imaging and Echocardiography in Patients With Hypertrophic Cardiomyopathy.

Hindieh W[1], Weissler-Snir A[1], Hammer H[1], Adler A[1], Rakowski H[1], Chan RH[2].

- 195 patients
- Echo and CMR within 6 months
- Maximal LVWT difference between echocardiography and CMR was 0.5 mm (95% confidence interval, -6.9, 7.8)
- In 90 patients (92.8%), echocardiography underestimated (n=32; 33.0%) or overestimated (n=58; 59.8%) maximal LVWT
- Under-estimation: focal LV hypertrophy (n=10; 10.3%) or poor acoustic windows (n=22; 22.7%)
- Over-estimation: inclusion RV (n=37; 38.1%), LV trabeculations (n=5; 5.2%),
  papillary muscle (n=3; 3.1%), and apical-septal bundle (n=1; 1.0%)
  imaging plane obliquity (n=7; 12.5%)

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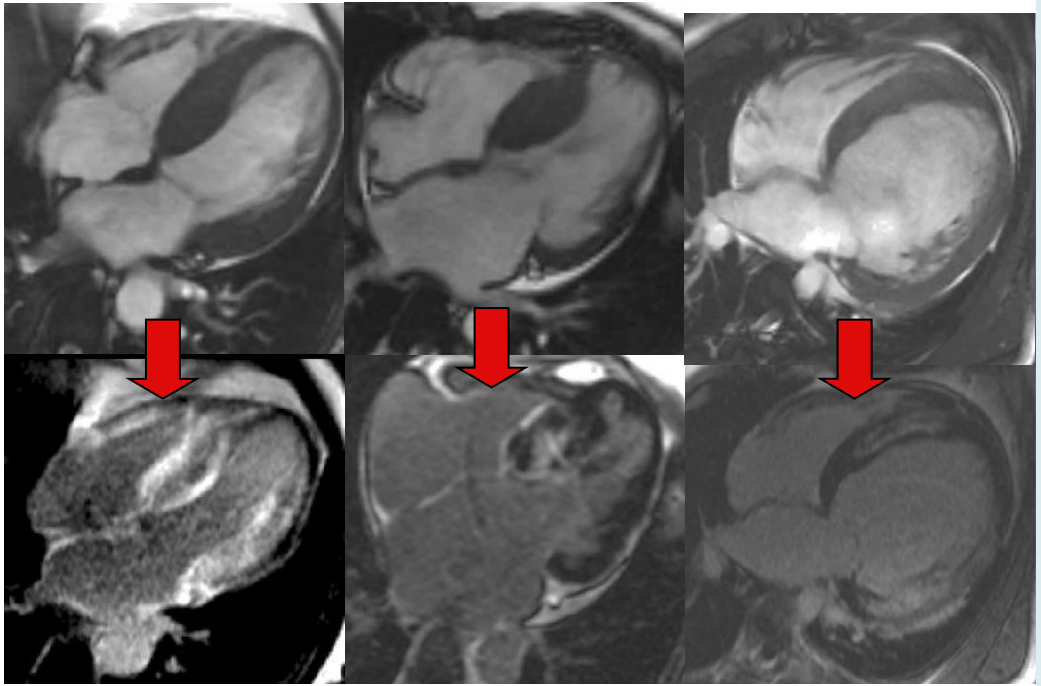
9/Differential diagnosis of LV hypertrophy



Amyloidosis HCM Fabry's

Table 5 Echocardiographic features that suggest specific aetiologies (modified from Rapezzi et al.[67])

Finding	Specific diseases to be considered
Increased interatrial septum thickness	Amyloidosis
Increased AV valve thickness	Amyloidosis; Anderson-Fabry disease
Increased RV free wall thickness	Amyloidosis, myocarditis, Anderson-Fabry disease, Noonan syndrome and related disorders
Mild to moderate pericardial effusion	Amyloidosis, myocarditis
Ground-glass appearance of ventricular myocardium on 2D echocardiography	Amyloidosis
Concentric LVH	Glycogen storage disease, Anderson-Fabry disease, PRKAG2 mutations
Extreme concentric LVH (wall thickness ≥30 mm)	Danon disease, Pompe disease
Global LV hypokinesia (with or without LV dilatation)	Mitochondrial disease, TTR-related amyloidosis, PRKAG2 mutations, Danon disease, myocarditis, advanced sarcomeric HCM, Anderson-Fabry disease
Right ventricular outflow tract obstruction	Noonan syndrome and associated disorders

11/Guidelines recommendation regarding hy

2014 ESC Guidelines on diag
management of hypertrophi

Recommendations for cardiovascular magnetic resonance evaluation in hypertrophic cardiomyopathy

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r Imaging

Recommendations	Class[a]	Level[b]	Ref.[c]
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	148,149
In the absence of contraindications, CMR with LGE is recommended in patients with suspected HCM who have inadequate echocardiographic windows, in order to confirm the diagnosis.	I	B	126,127
In the absence of contraindications, 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	124,126,127,130 136,138–143
CMR with LGE imaging should be considered in patients with suspected apical hypertrophy or aneurysm.	IIa	C	127,129
CMR with LGE imaging should be considered in patients with suspected cardiac amyloidosis.	IIa	C	22,147
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	150,151

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1/ Role of CMR according to the 2010 task force criteria (Marcus et al, Circulation 2010)

Original Task Force Criteria	Revised Task Force Criteria
I. Global or regional dysfunction and structural alterations*	
Major	
• Severe dilatation and reduction of RV ejection fraction with no (or only mild) LV impairment • Localized RV aneurysms (akinetic or dyskinetic areas with diastolic bulging) • Severe segmental dilatation of the RV	**By 2D echo:** • Regional RV akinesia, dyskinesia, or aneurysm • *and* 1 of the following (end diastole): — PLAX RVOT ≥32 mm (corrected for body size [PLAX/BSA] ≥19 mm/m^2) — PSAX RVOT ≥36 mm (corrected for body size [PSAX/BSA] ≥21 mm/m^2) — *or* fractional area change ≤33% **By MRI:** • Regional RV akinesia or dyskinesia or dyssynchronous RV contraction • *and* 1 of the following: — Ratio of RV end-diastolic volume to BSA ≥110 mL/m^2 (male) or ≥100 mL/m^2 (female) — *or* RV ejection fraction ≤40% **By RV angiography:** • Regional RV akinesia, dyskinesia, or aneurysm

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1/ Role of CMR according to the 2010 task force criteria (Marcus et al, Circulation 2010)

Minor

- Mild global RV dilatation and/or ejection fraction reduction with normal LV
- Mild segmental dilatation of the RV
- Regional RV hypokinesia

By 2D echo:

- Regional RV akinesia or dyskinesia
- *and* 1 of the following (end diastole):
  - PLAX RVOT ≥29 to <32 mm (corrected for body size [PLAX/BSA] ≥16 to <19 mm/m^2)
  - PSAX RVOT ≥32 to <36 mm (corrected for body size [PSAX/BSA] ≥18 to <21 mm/m^2)
  - *or* fractional area change >33% to ≤40%

By MRI:

- Regional RV akinesia or dyskinesia or dyssynchronous RV contraction
- *and* 1 of the following:
  - Ratio of RV end-diastolic volume to BSA ≥100 to <110 mL/m^2 (male) or ≥90 to <100 mL/m^2 (female)
  - *or* RV ejection fraction >40% to ≤45%

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2/ Appropriate ways to assess right ventricular end-diastolic volume (RVEDV) and right ventricular ejection fraction (RVEF) and wall motion abnormalities

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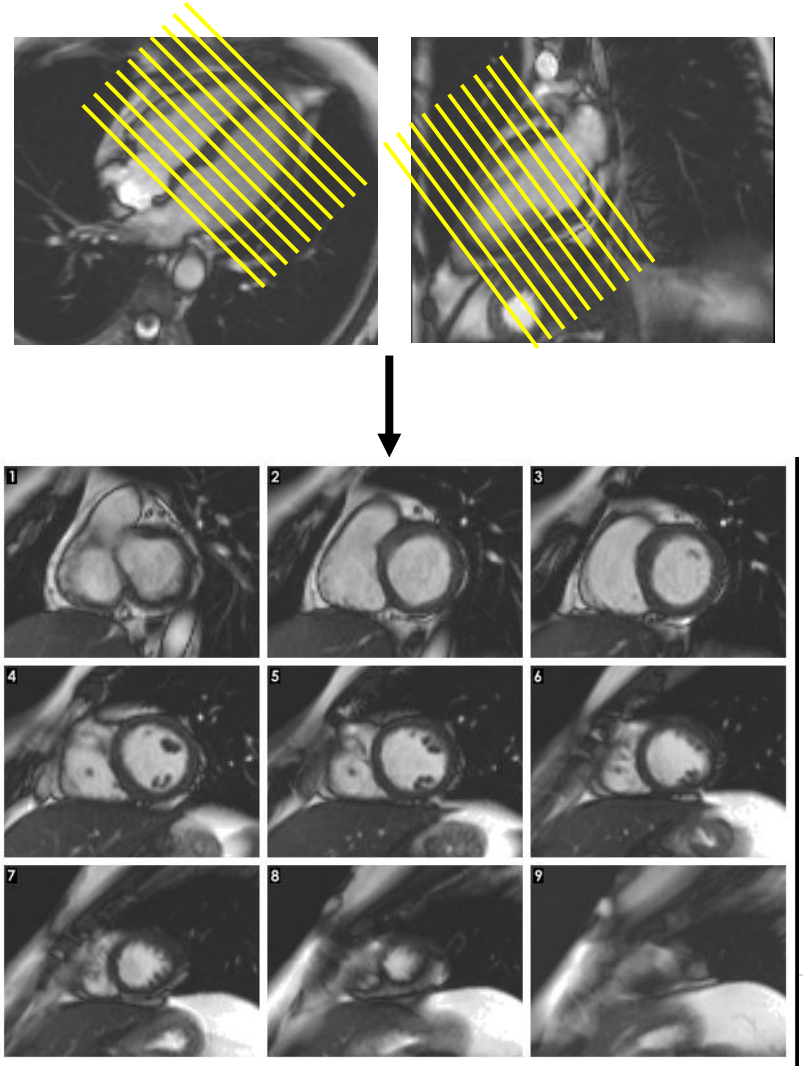
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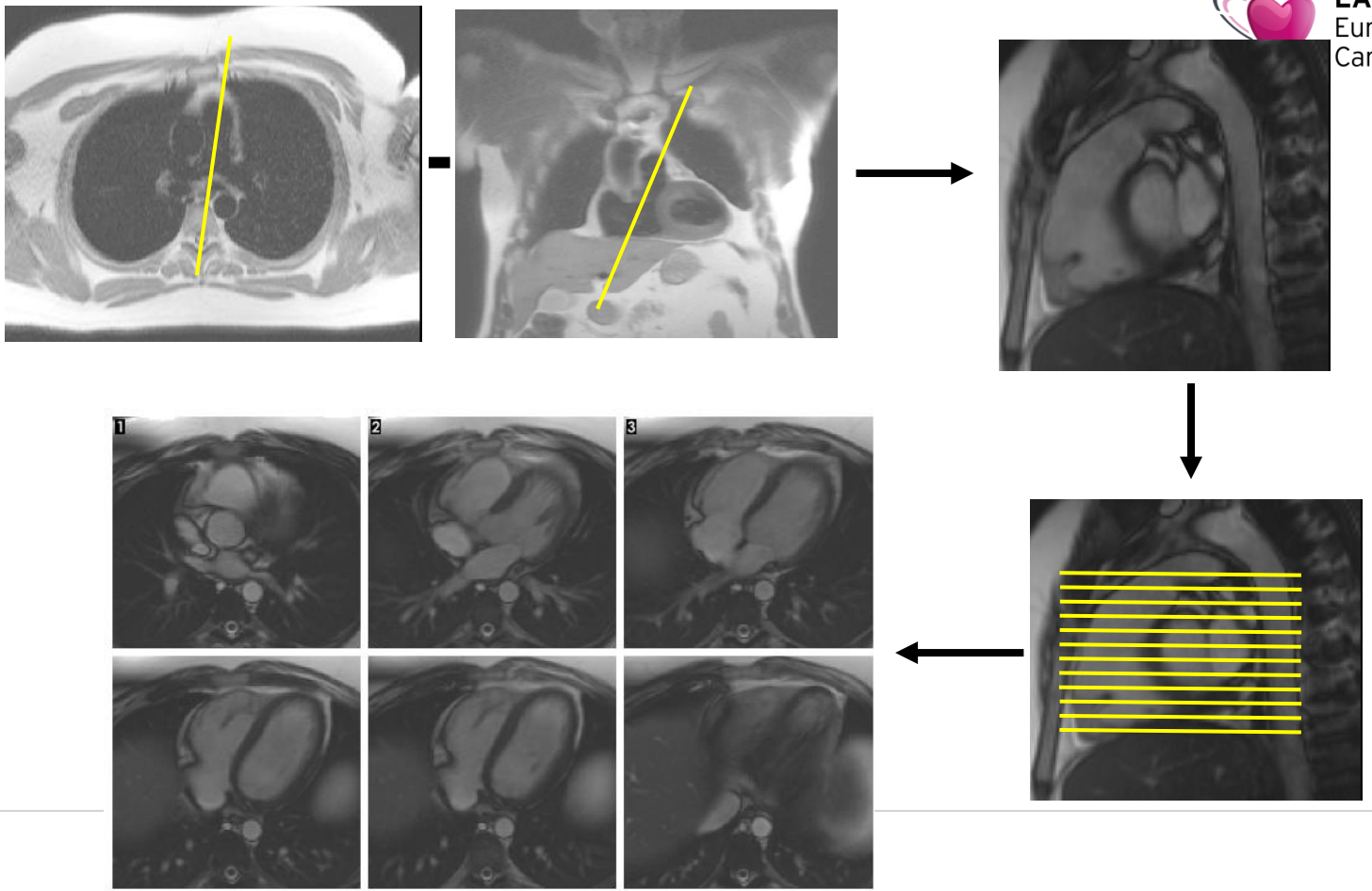
Additional RV Images



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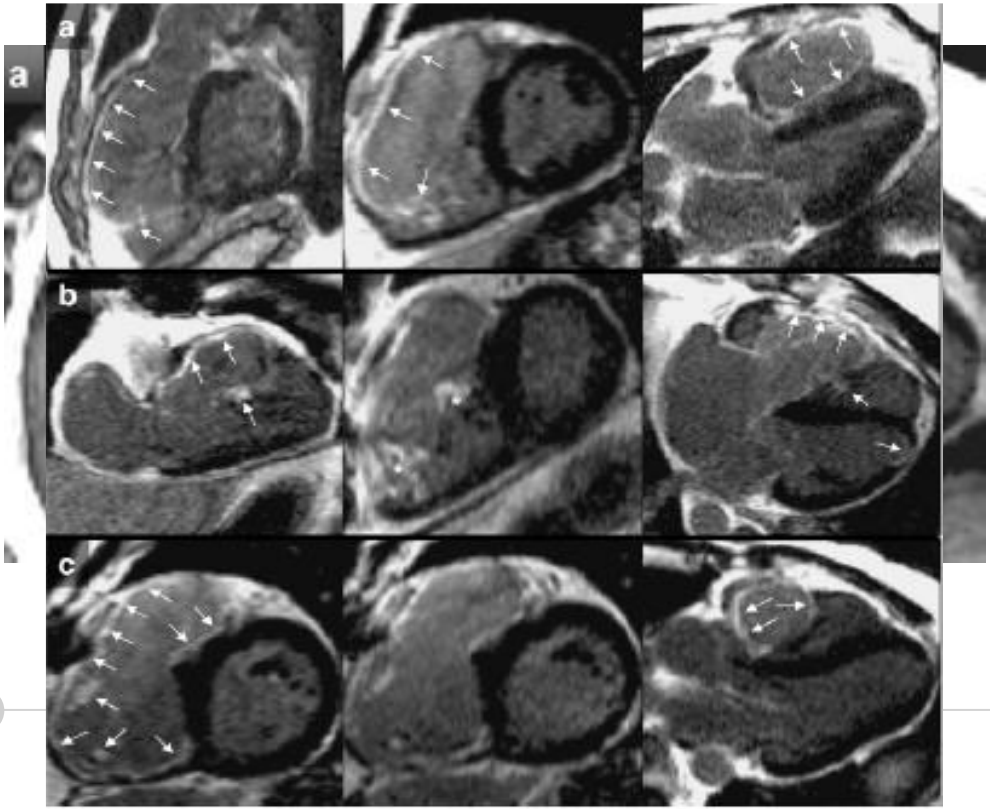
4/ Importance, pitfalls, challenges of RV LGE



- Thin structure
- TI for RV (? same for LV)
- Prominent epicardial fat

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ARVC- Cine and LGE

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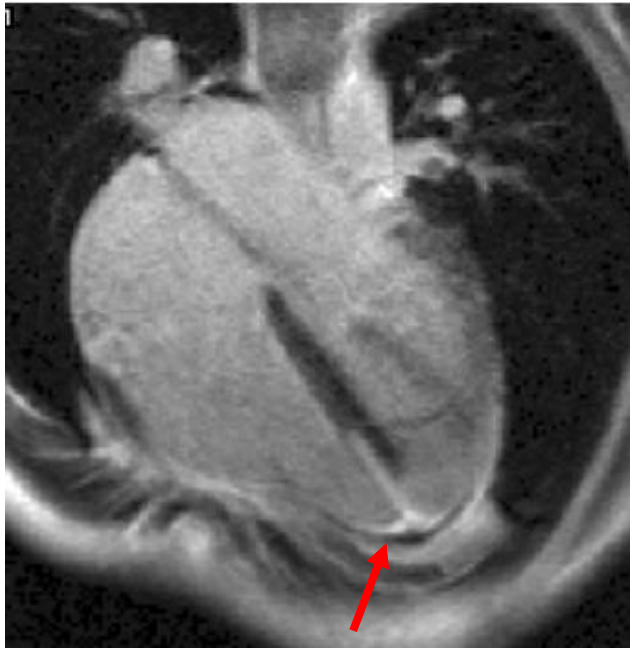

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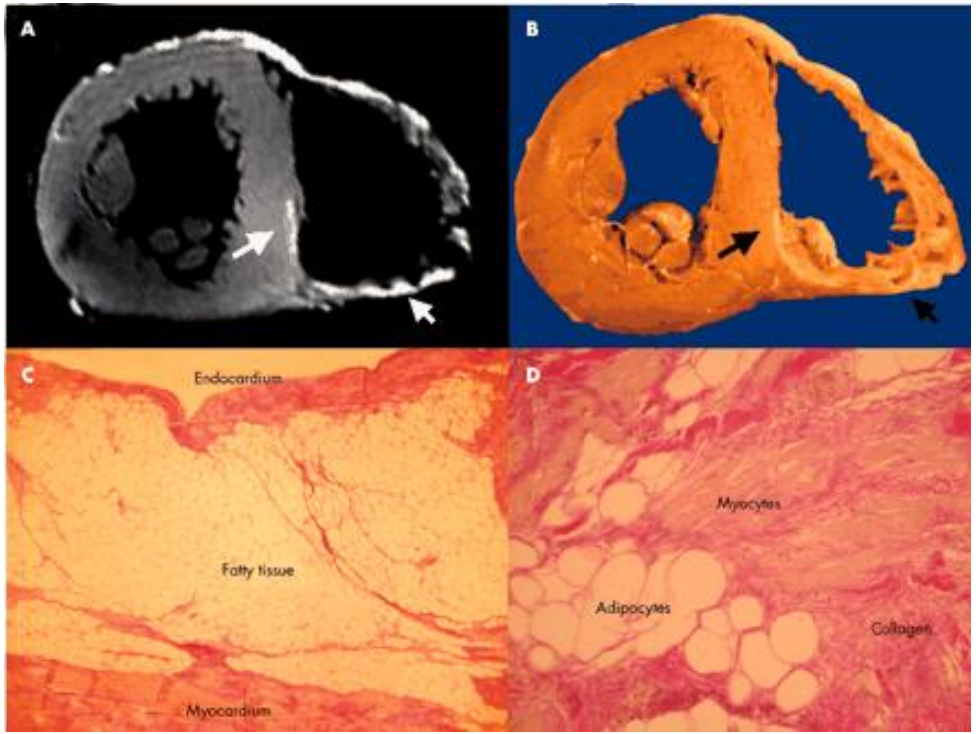

John AS, Heart 2004

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5/ Differential diagnosis of RV enlargement

Arrhythmogenic right ventricular cardiomyopathy mimics: role of cardiovascular magnetic resonance

Giovanni Quarta[1], Syed I Husain[1], Andrew S Flett[1], Daniel M Sado[1,2], Charles Y Chao[1], Mariá T Tomé Esteban[1], William J McKenna[1,2], Antonios Pantazis[1] and James C Moon[1,2*]

JCMR 2013

1. Displacement of the heart (pectus, partial absence pericardium, etc)
2. RV overload (volume/pressure) (ASD, anomalous venous return, pulmonary hypertension)
3. RV scarring (RV infarction, RV involvement in cardiac sarcoidosis

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Clinical Case

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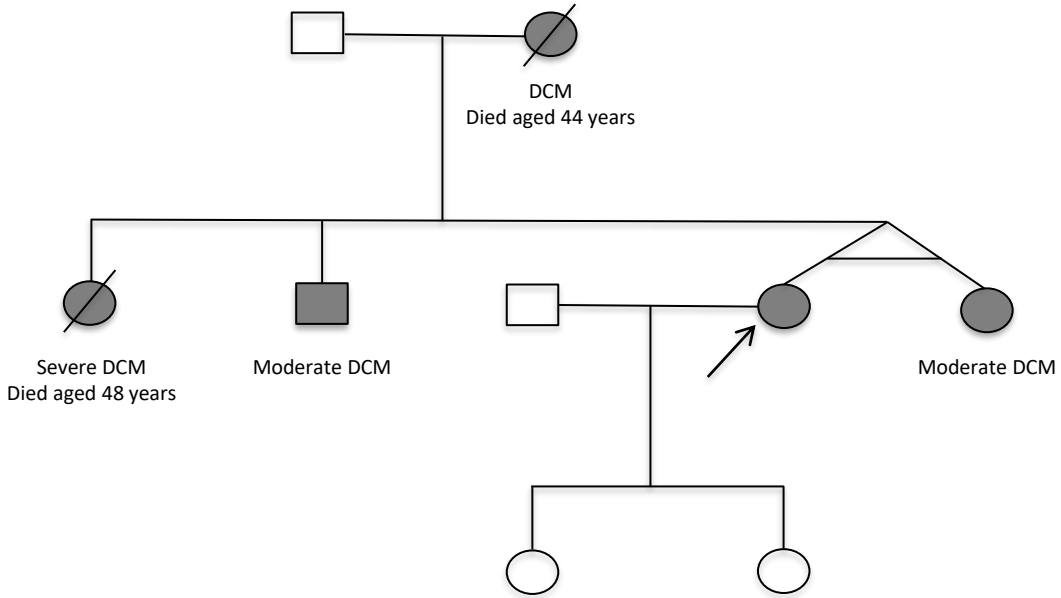
- **36 year old asymptomatic woman**



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Clinical Case

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- **ECG:**
  - Normal
- **ECHO:**
  - EF = 35%, LVIDd = 57mm
- **Clinical course:**
  - Stable echo for 7 years
  - **Palpitation** => 24 hour tape

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Clinical Case

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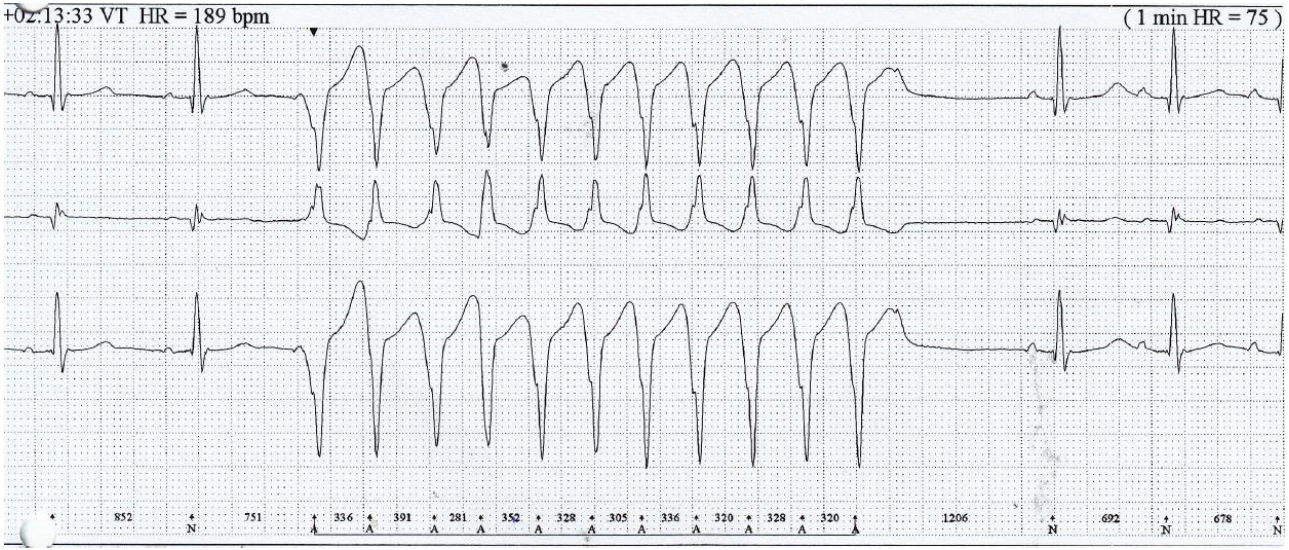
- **24-hour tape:**



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LV Function

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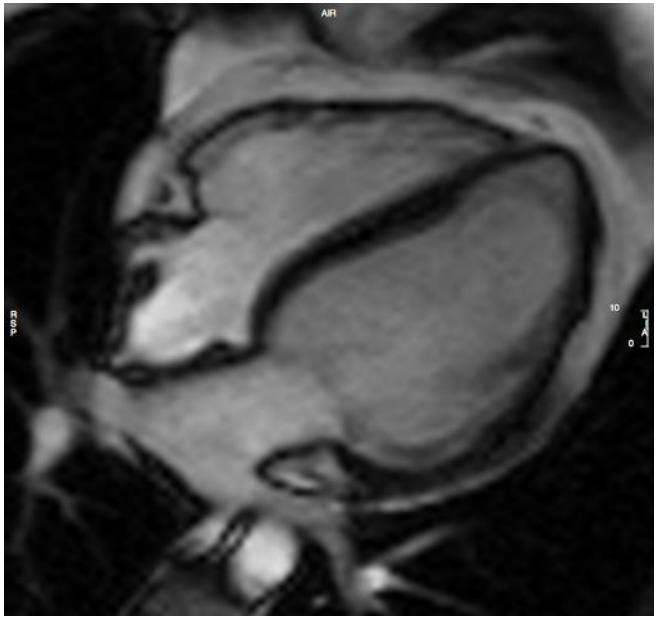
Short axis
SSFP cine



4 chamber
SSFP cine

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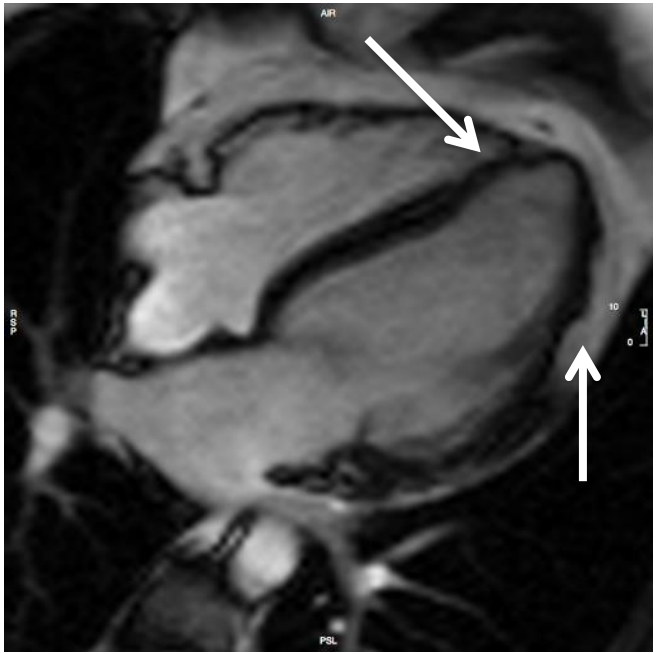
LV Function



4 chamber
SSFP cine

Tissue Characterisation

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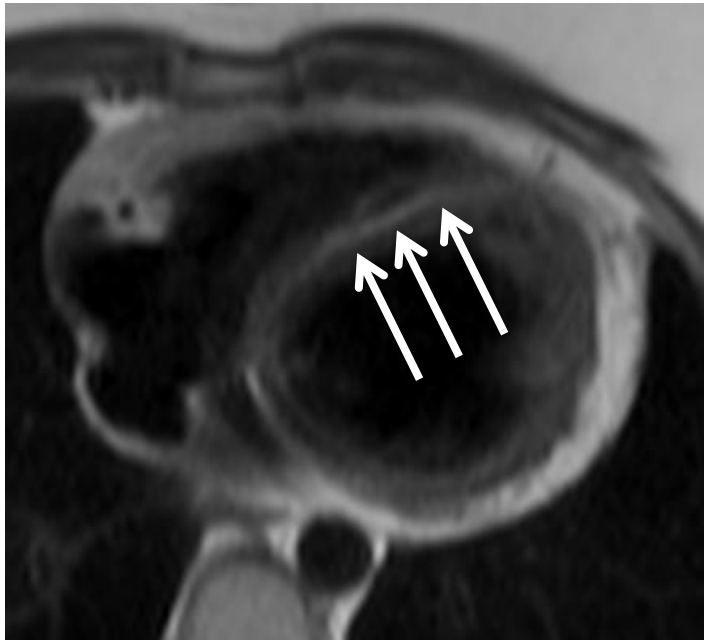
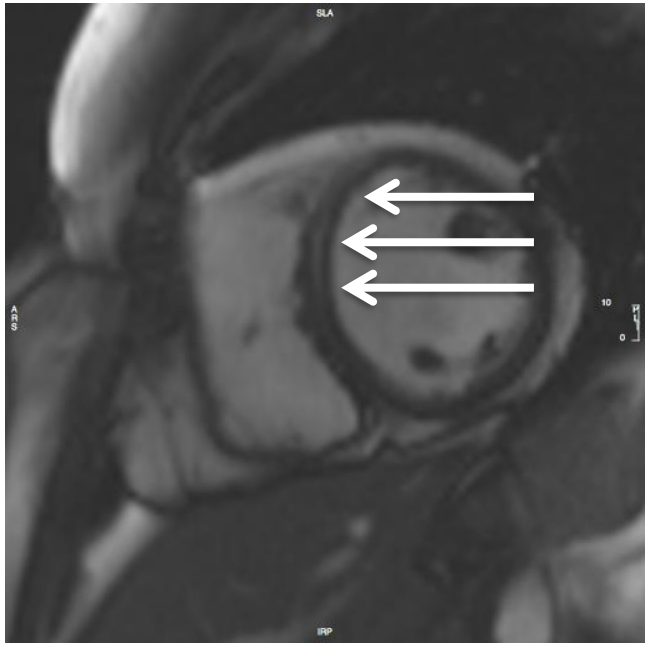


Axial HASTE
T1



Short axis SSFP cine
T2 > T1

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Tissue Characterisation

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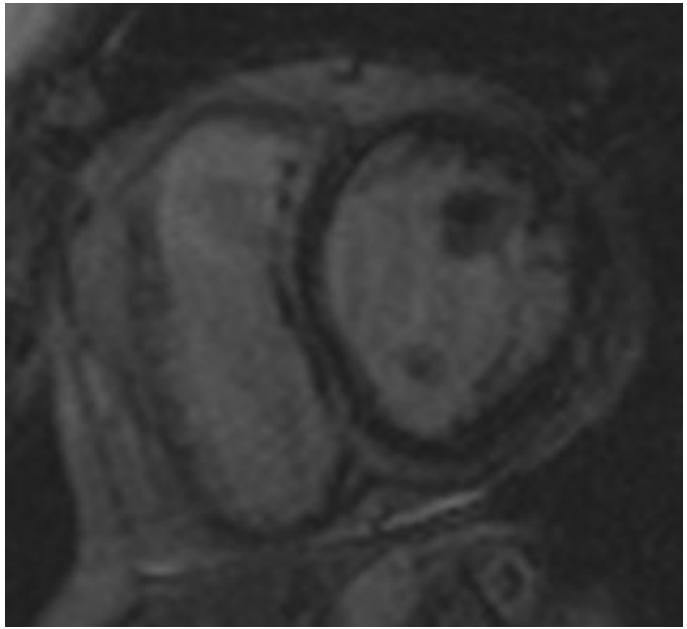
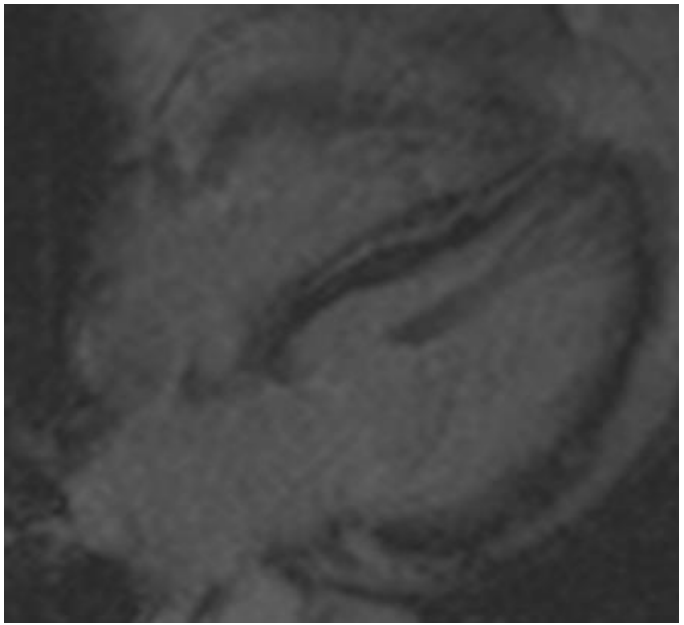


Short axis
LGE



4-Chamber
LGE

Diagnosis: ALVC

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Learning Points

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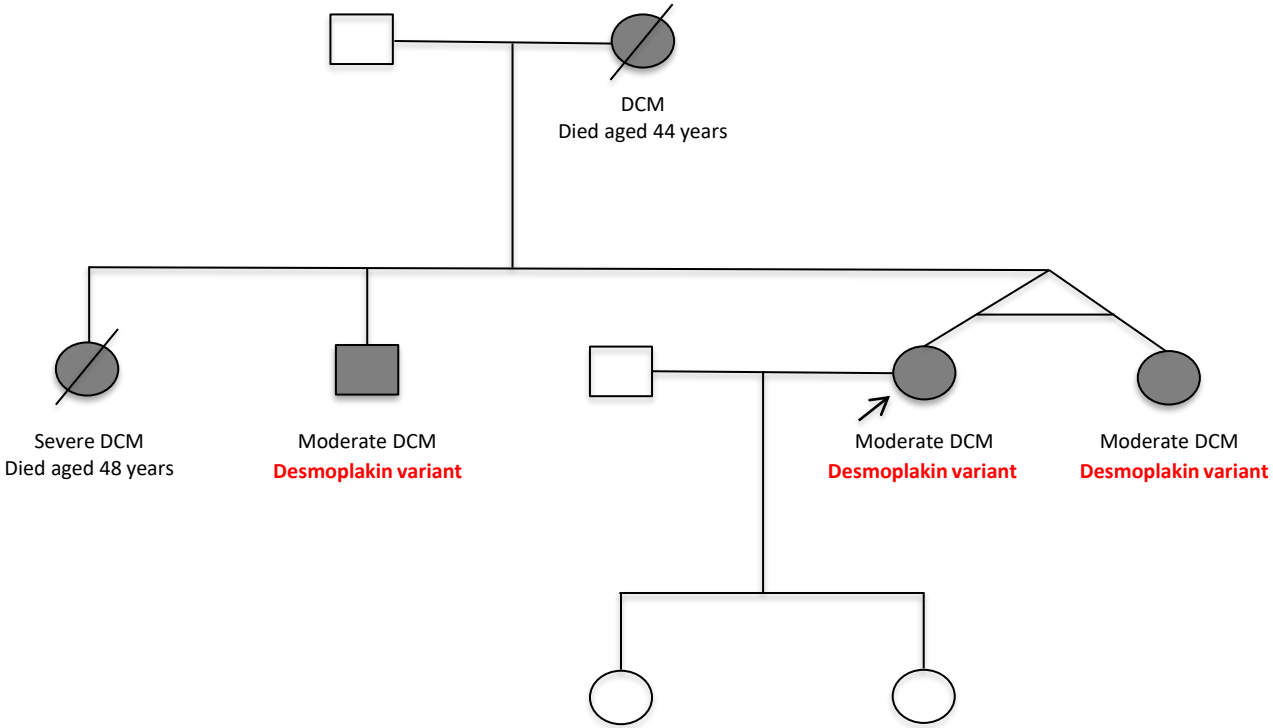

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Learning Points

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CMR:

- **Identified underlying cardiomyopathic process**
  - Not achieved with preceding echocardiograms
- **Influenced patient management**
  - Identified substrate for malignant arrhythmia
  - Prompted ICD implantation
- **Influenced family screening[1]**
  - Desmoplakin gene

[1] Sen-Chowdhry et al. J Am Coll Cardiol. 2008; 52: 2175 – 87.

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ESC European Society of Cardiology

European Heart Journal (2022) **00**, 1–130
https://doi.org/10.1093/eurheartj/ehac262

ESC GUIDELINES

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2022 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

Developed by the task force for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death of the European Society of Cardiology (ESC)

Endorsed by the Association for European Paediatric and Congenital Cardiology (AEPC)

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. CMR with LGE is recommended in hypertrophic cardiomyopathy (HCM) patients for diagnostic workup. Genetic counseling and testing are also recommended. In a first-degree relative of a patient with HCM, ECG and echocardiogram are recommended. ICD implantation should be considered in HCM patients with an intermediate 5-year risk of SCD, and with: a) significant LGE at CMR; or b) LVEF <50%; or c) abnormal blood pressure response during exercise test; or d) LV apical aneurysm; or e) presence of sarcomeric pathogenic mutation.

In patients with suspected arrhythmogenic right ventricular cardiomyopathy (ARVC), CMR is recommended. In patients with suspected or definite diagnosis of ARVC, genetic counseling and testing are recommended. ICD implantation should be considered in symptomatic patients with definite ARVC, moderate right or left ventricular dysfunction, and either nonsustained VT or inducibility of SMVT at EP study.

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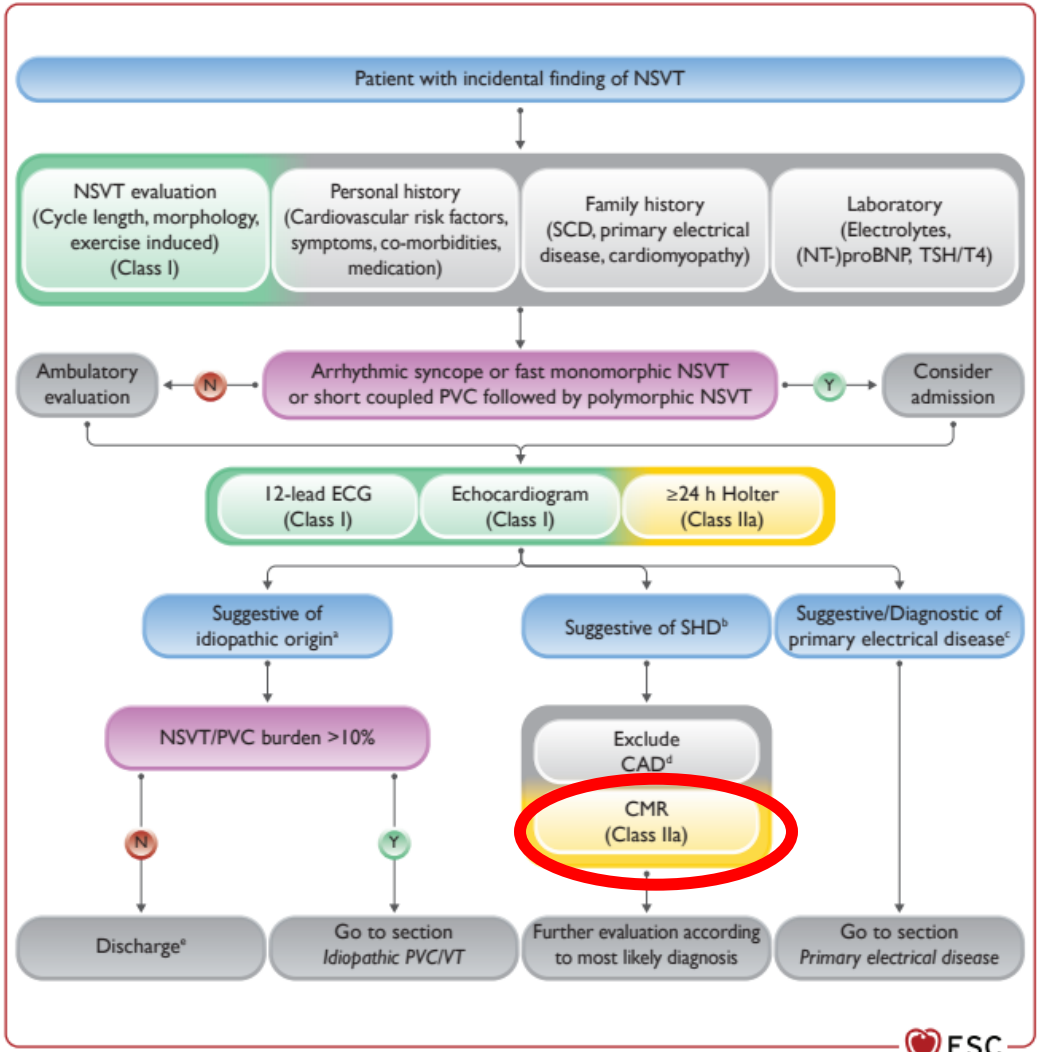
SVMT=Suspected monomorphic VT



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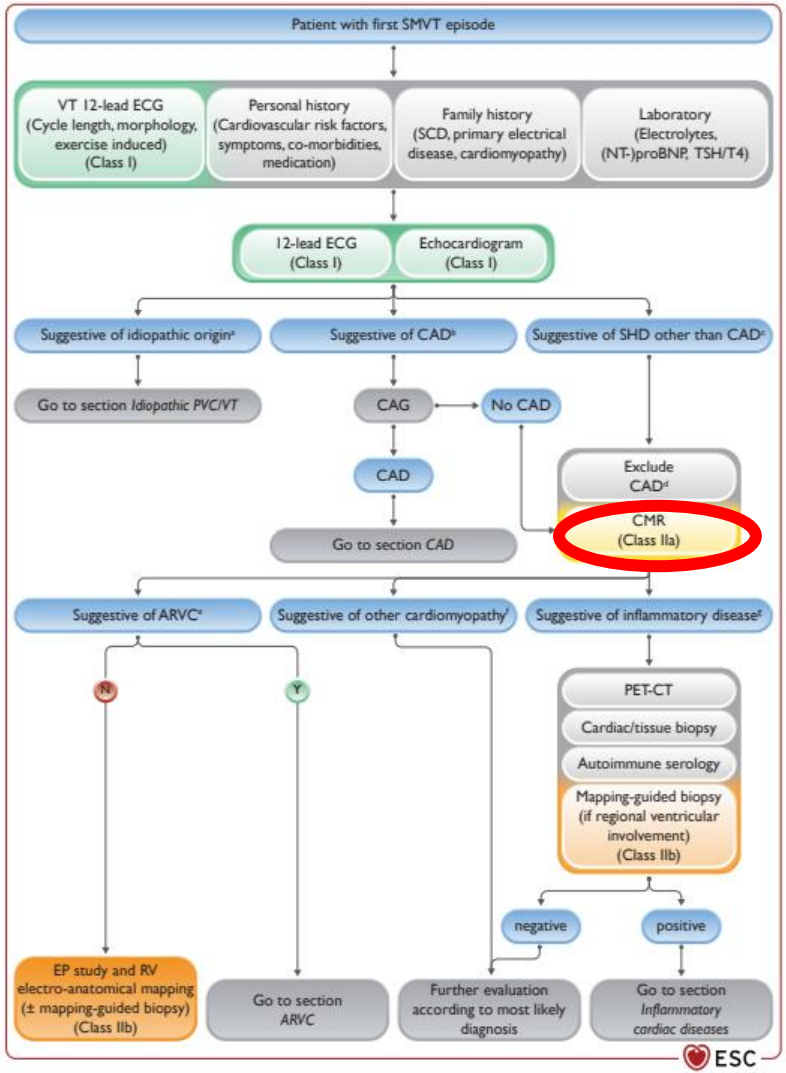

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