2019 VT symposium
"Session title: Hypertrophic cardiomyopathy and VT/VF"

Imaging for Diagnosis and SCD Risk

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- Imaging for diagnosis of HCMP
- Imaging for SCD risk assessment in patients with HCMP

European Heart Journal (2014) **35**, 2733–2779
doi:10.1093/eurheartj/ehu284

ESC GUIDELINES

2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy

The Task Force for the Diagnosis and Management of Hypertrophic Cardiomyopathy of the European Society of Cardiology (ESC)

Authors/Task Force members: Perry M. Elliott* (Chairperson) (UK) Aris Anastasakis (Greece), Michael A. Borger (Germany), Martin Borggrefe (Germany), Franco Cecchi (Italy), Philippe Charron (France), Albert Alain Hagege (France), Antoine Lafont (France), Giuseppe Limongelli (Italy), Heiko Mahrholdt (Germany), William J. McKenna (UK), Jens Mogensen (Denmark), Petros Nihoyannopoulos (UK), Stefano Nistri (Italy), Petronella G. Pieper (Netherlands), Burkert Pieske (Austria), Claudio Rapezzi (Italy), Frans H. Rutten (Netherlands), Christoph Tillmanns (Germany), Hugh Watkins (UK).

Imaging for diagnosis of HCMP

Traditional concept of HCMP

- The presence of ***increased LV wall thickness*** that is not solely explained by abnormal loading conditions like hypertension, aortic stenosis, and so on.

- First to diagnose HCM clinically in the 1960s by ***Dr. Braunwald***

- ***m/c genetic cardiomyopathy***
  - Prevalence of **1:500 from the CARDIA study**
  - Typically inherited in AD, but variable penetrance, expressivity

Maron BJ et al. Circulation 1995;92:785
Maron BJ et al. JAMA 2002;287:1308
Elliott PM et al. EHJ 2014;35:2733

Traditional concept of HCMP

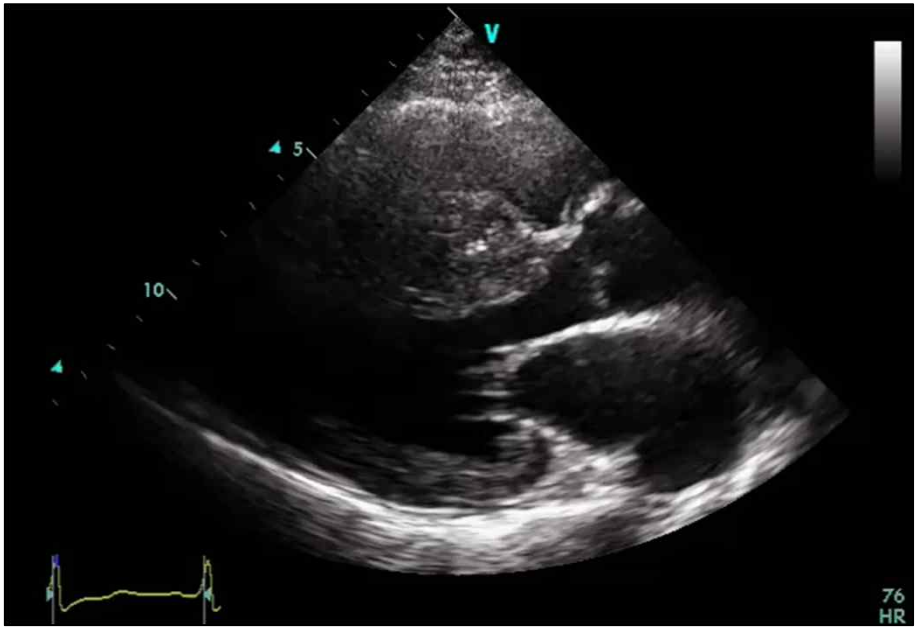
- ***Exclusion* diagnosis**
  - Secondary causes of LVH should be ruled out
    : Systemic HT, valvular/subvalvular AS, infiltrative CMP

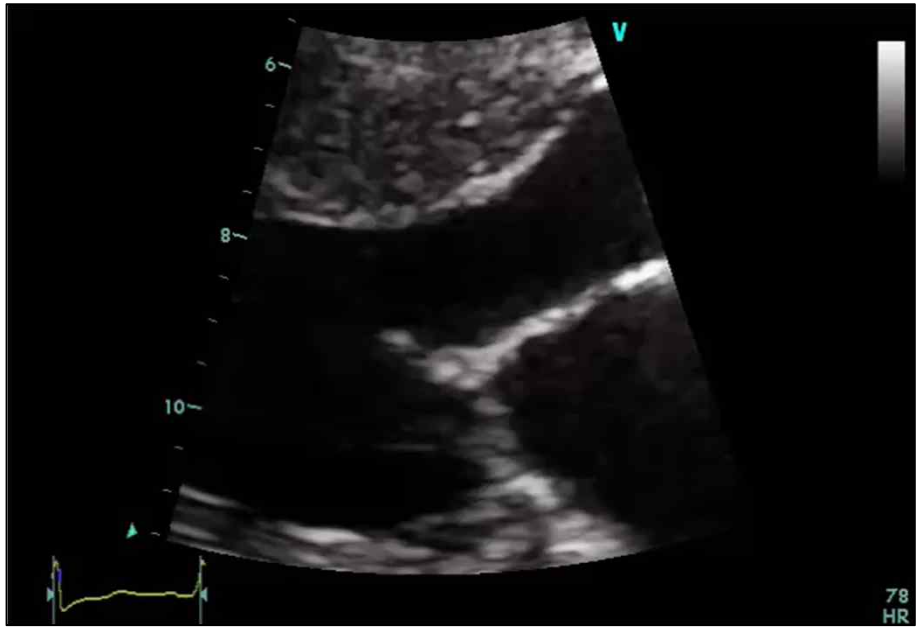
- ***Morphologic* diagnosis**

  primarily by *transthoracic echocardiography*
  - Increased LV wall thickness ≥15mm
  - Increased LV wall thickness ≥13mm in first-degree family members of patients with unequivocal HCM

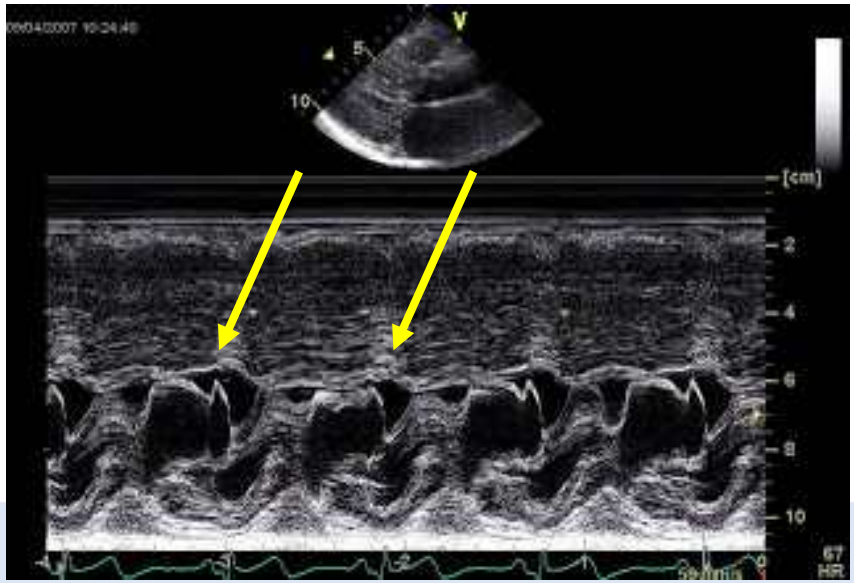
Maron BJ et al. JAMA 2002;287:1308
Elliott PM et al. EHJ 2014;35:2733

Traditional diagnosis - EchoCG

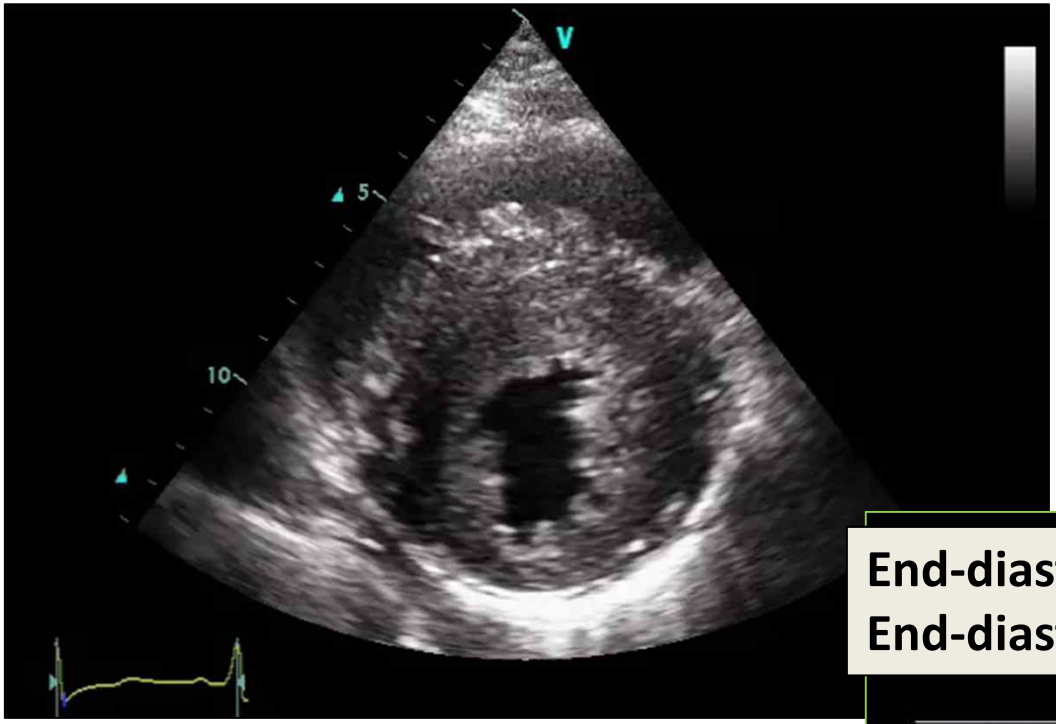




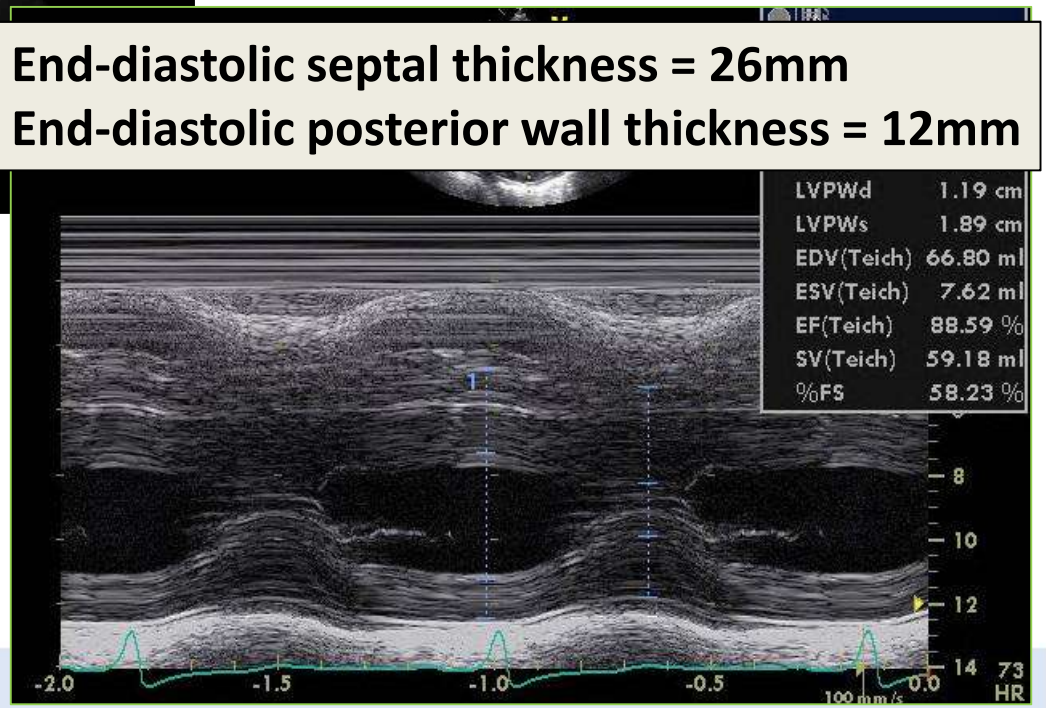
- Asymmetric Septal hypertrophy
- Thickness = 28mm
- SAM



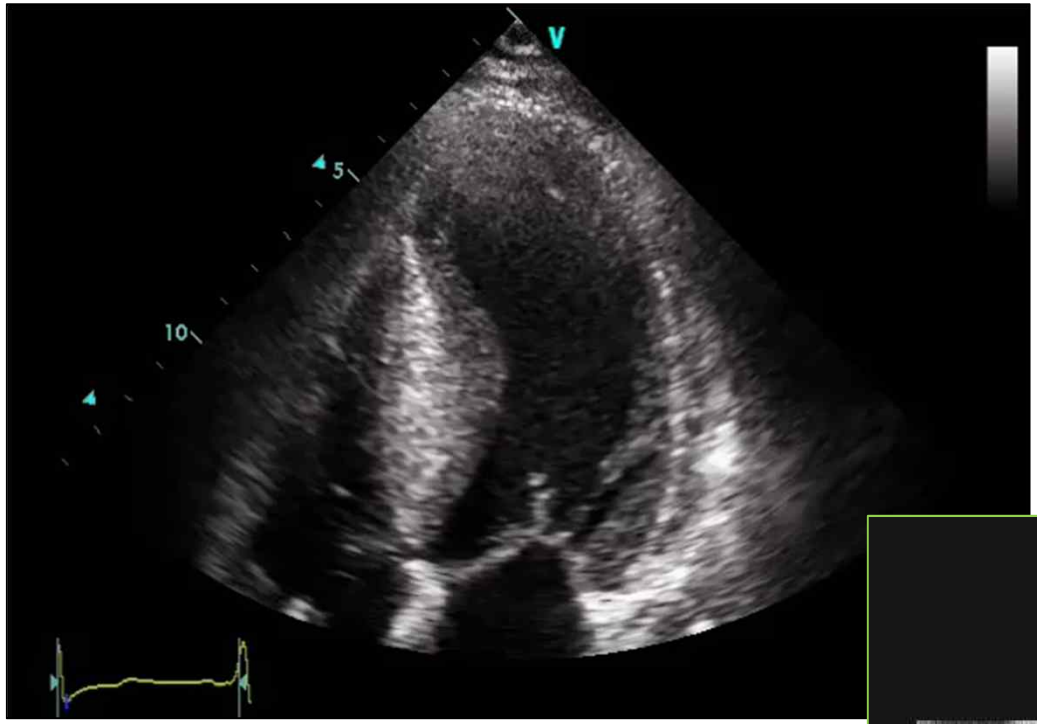
Traditional diagnosis - EchoCG

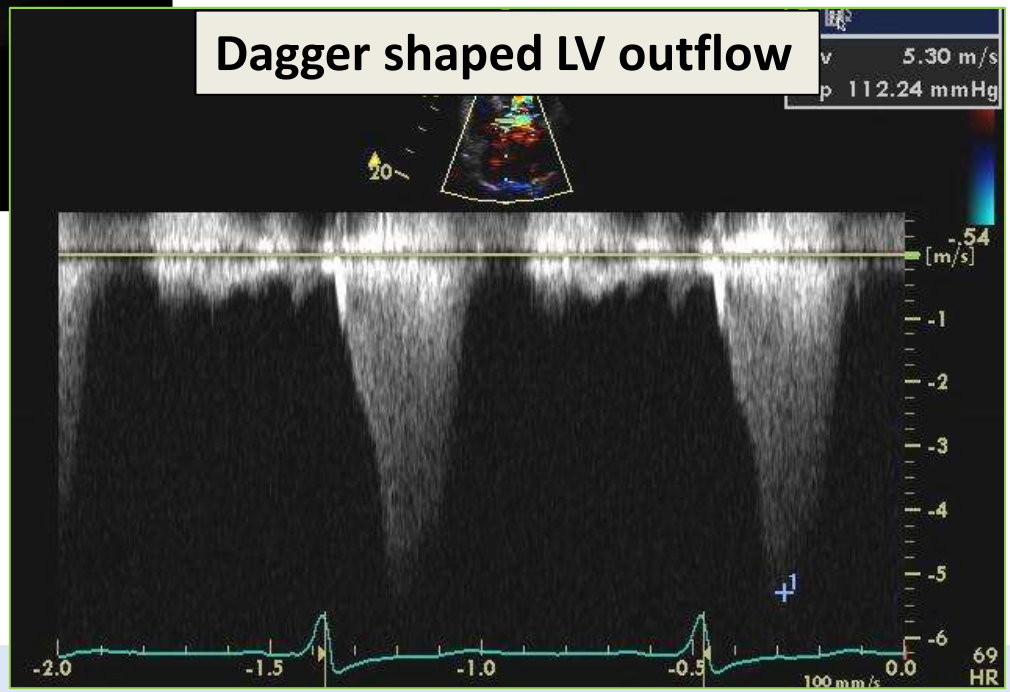


End-diastolic septal thickness = 26mm
End-diastolic posterior wall thickness = 12mm

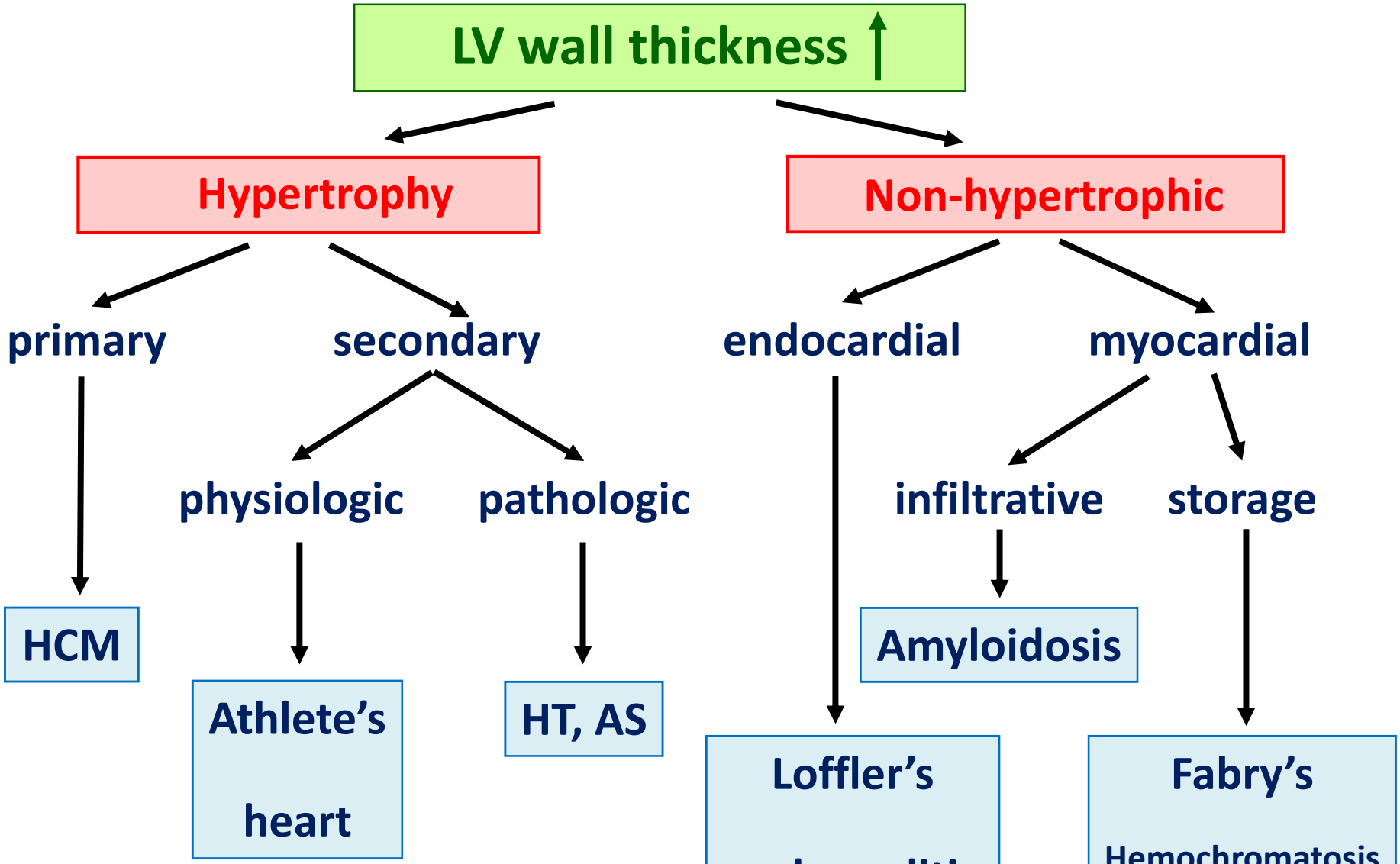


Traditional diagnosis - EchoCG





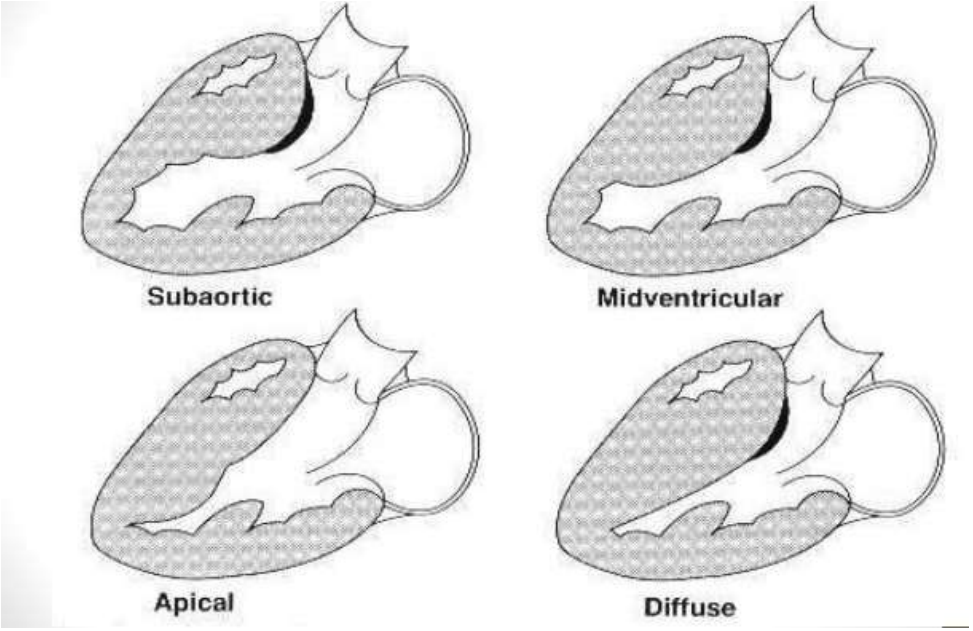
Differential diagnosis of increased LV thickness



Courtesy of HK Kim

EchoCG in diagnosis of HCMP

- Demonstrate severity and distribution of **LVH**

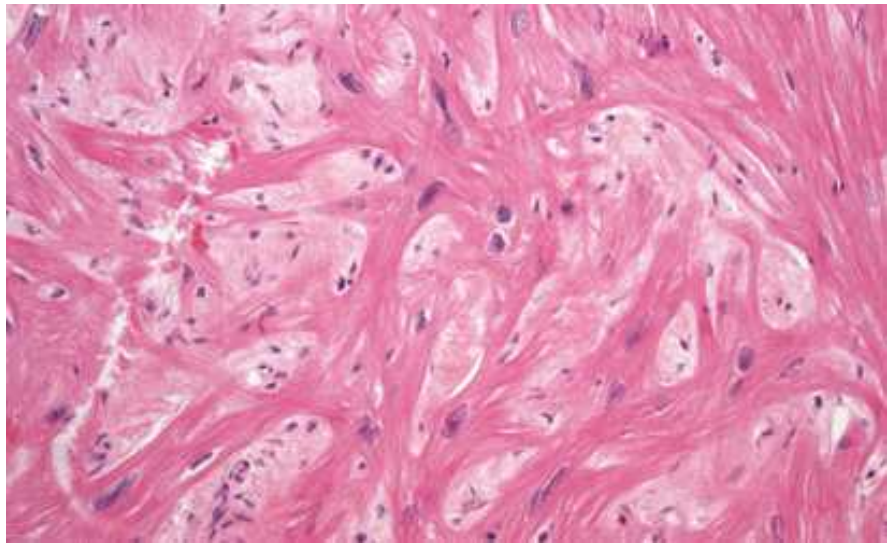


- Mitral valve abnormality : SAM, MR, MV abnormality
- Assessment of obstruction
- Evaluate **diastolic dysfunction**

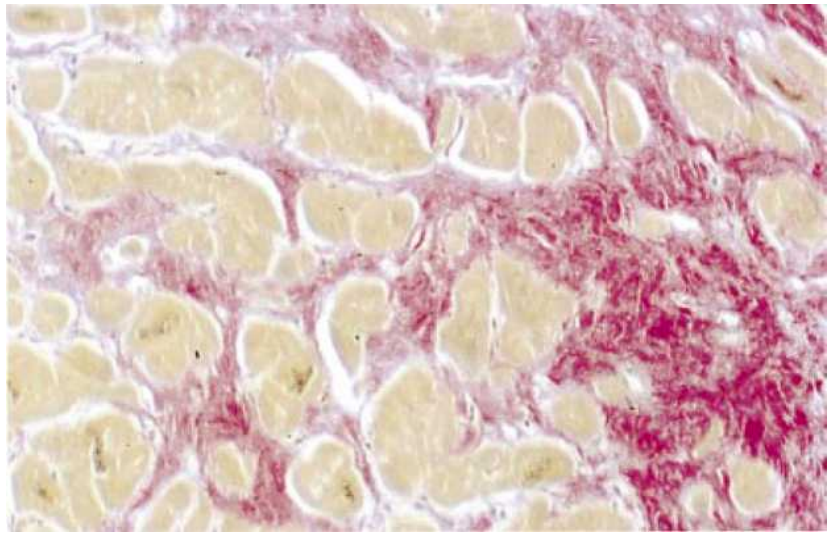
Bos JM et al. JACC 2009;54:201
Geske JB et al. JACC HF 2018;6:364

Traditional concept of HCMP

- ***Histologic diagnosis***
  - Myocardial fiber disarray with interstitial fibrosis, ECM expansion



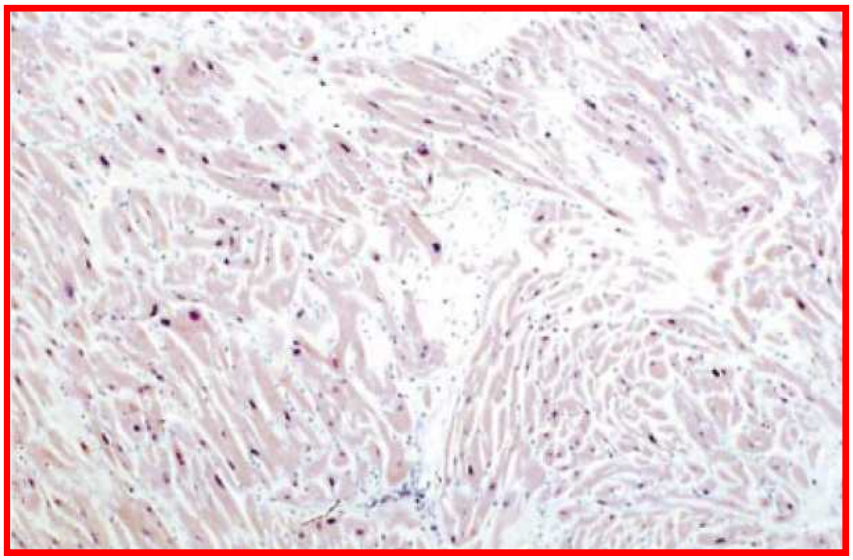
Myocardial fiber disarray

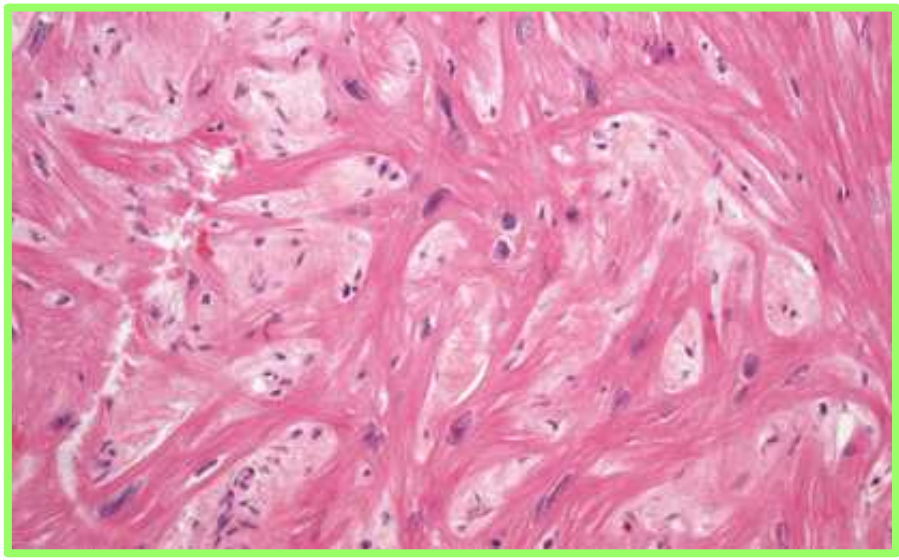


Replacement & Interstitial fibrosis

Maron BJ et al. JAMA 2002;287:1308
Gersh BJ et al. JACC 2011;58:e212
Elliott PM et al. EHJ 2014;35:2733

Limitations of histologic diagnosis





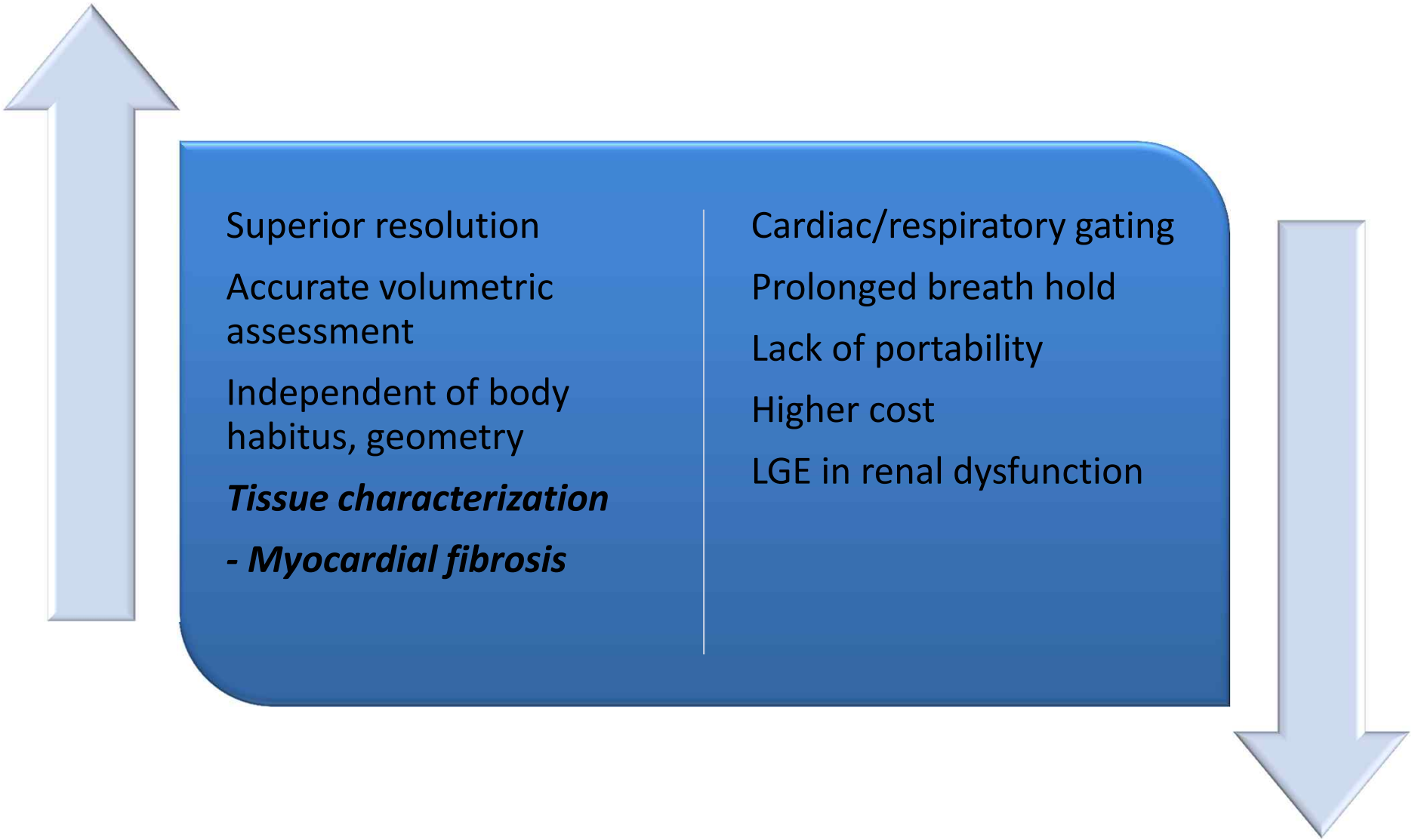
- **Not specific for sarcomeric disease !**

  - Anderson-Fabry disease — *Cardiovasc Pathol. 2010;19:293-301*
  - Noonan syndrome — *Br Heart J. 1992;68:586-588*
  - Friedreich ataxia — *J Clin Invest. 2002;109:357-362*

  * in trabeculations and RV insertion points in normal heart

- **No direct clinical surrogate for myocyte disarray**

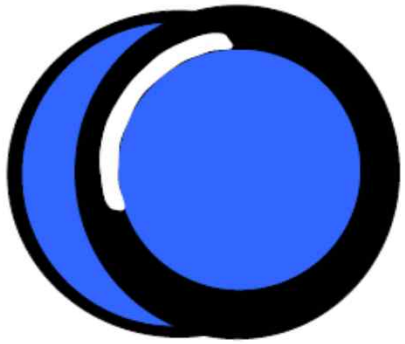
Instead, using LGE on CMR



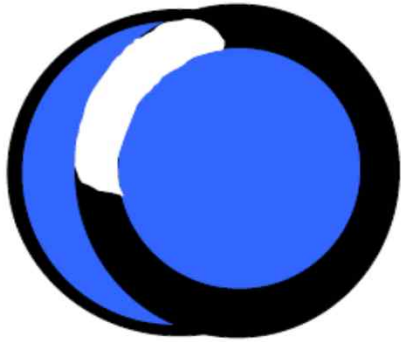
Differential diagnosis by LGE pattern

Ischemic pattern

A. Subendocardial infarct

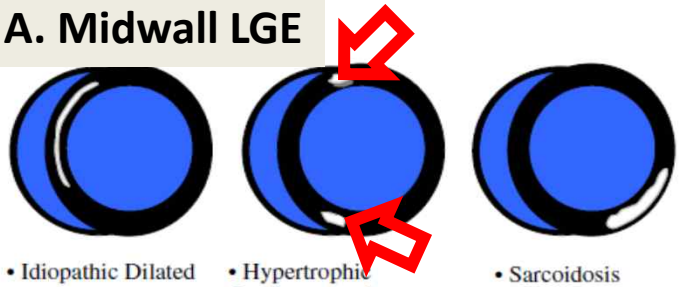


B. Transmural infarct



Non-ischemic pattern

A. Midwall LGE

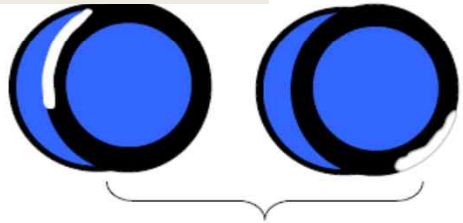


- Idiopathic Dilated Cardiomyopathy
- Myocarditis

- Hypertrophic Cardiomyopathy
- Right ventricular pressure overload (e.g. congenital heart disease, pulmonary HTN)

- Sarcoidosis
- Myocarditis
- Anderson-Fabry's disease
- Chagas' disease

B. Epicardial LGE



- Sarcoidosis, Myocarditis, Anderson-Fabry's disease, Chagas' disease

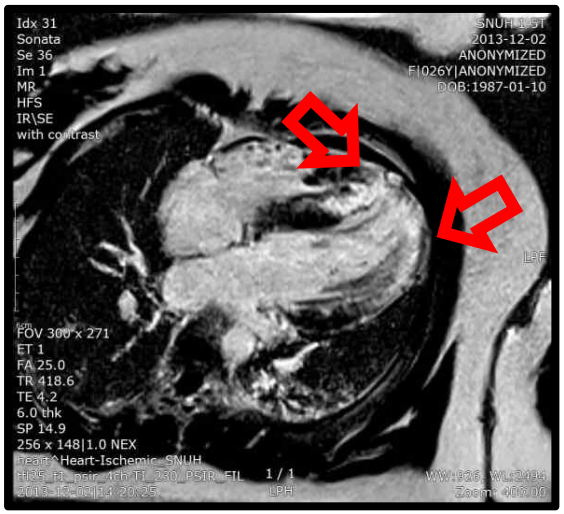
C. Global subendocardial LGE

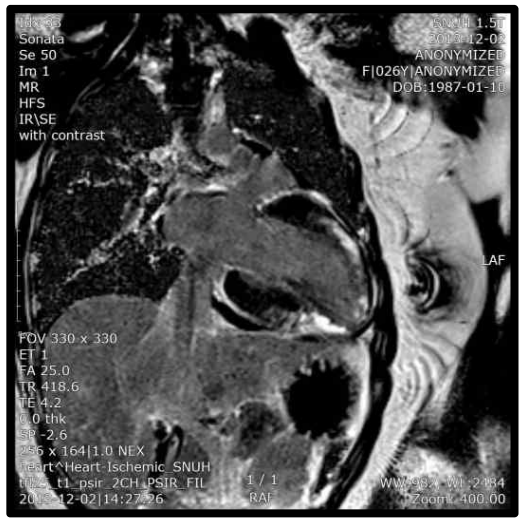
- Amyloidosis, Systemic Sclerosis, Post cardiac transplantation

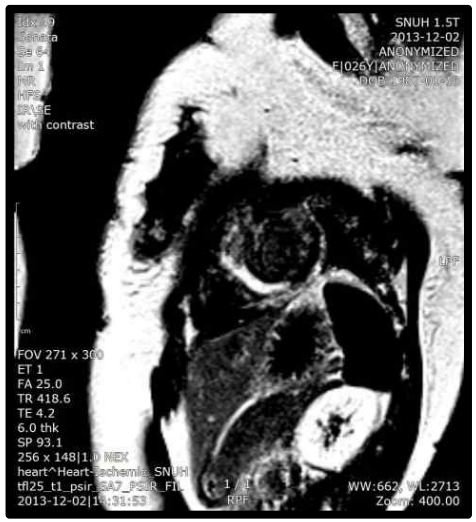
Shah DJ and Kim RJ et al. Clinical MRI. 3rd Edition, NY, 2006

Not easy to diagnose by LGE pattern

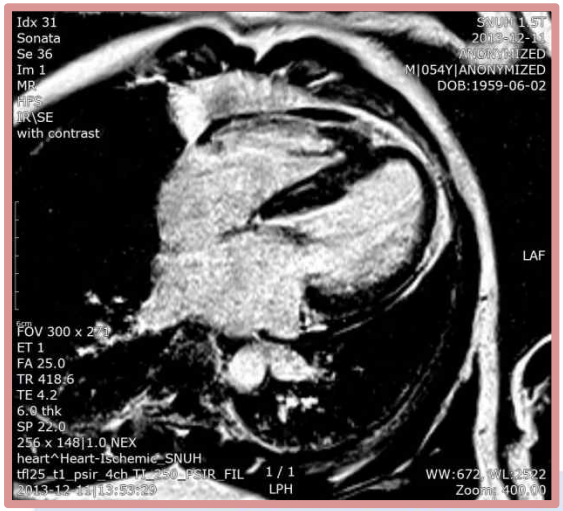
- **23 year-old female**

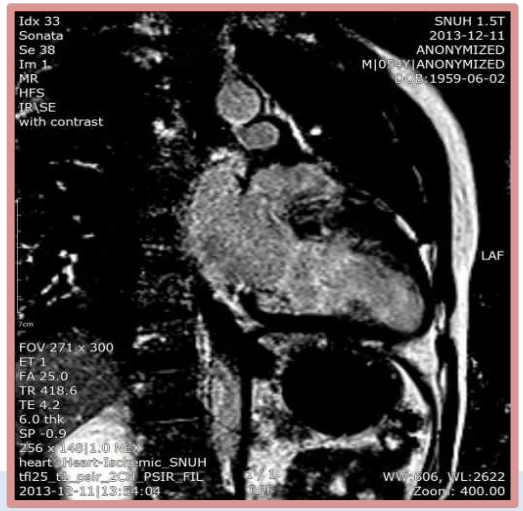


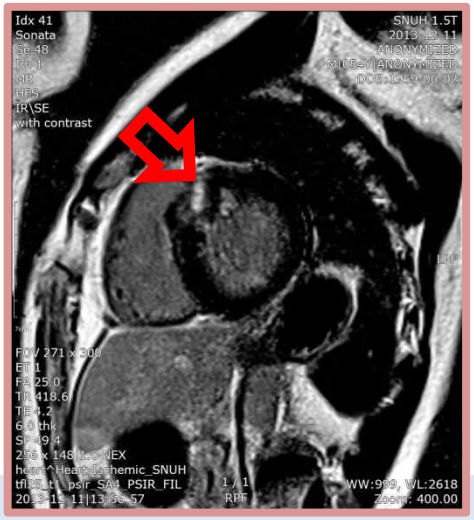




- **46 year-old male**

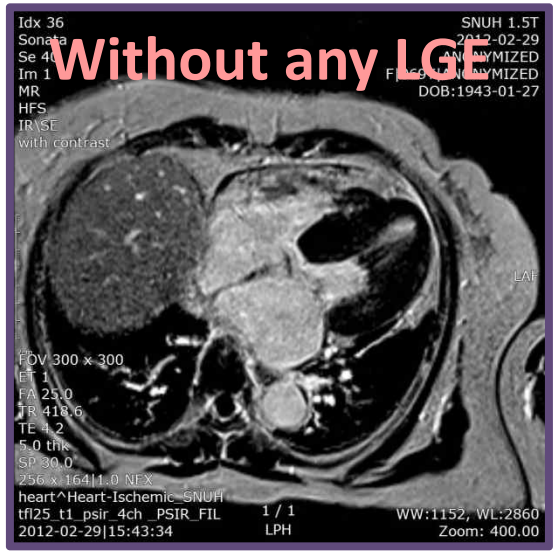


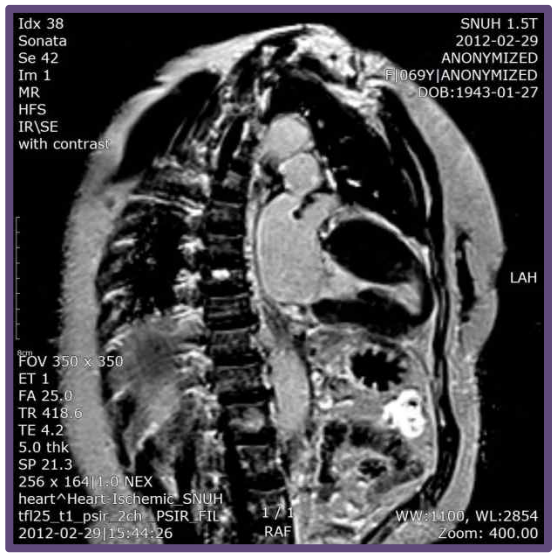


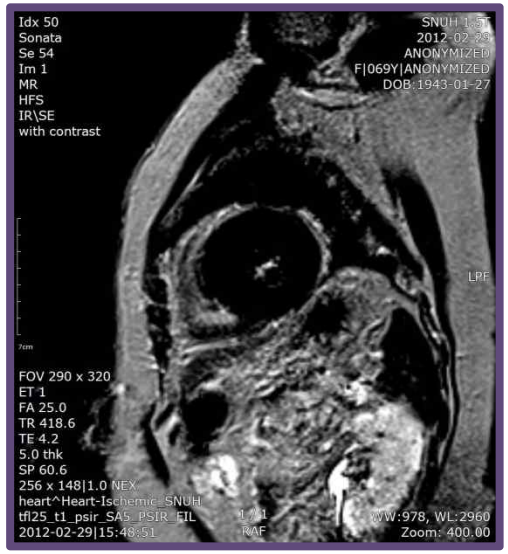


Not easy to diagnose by LGE pattern

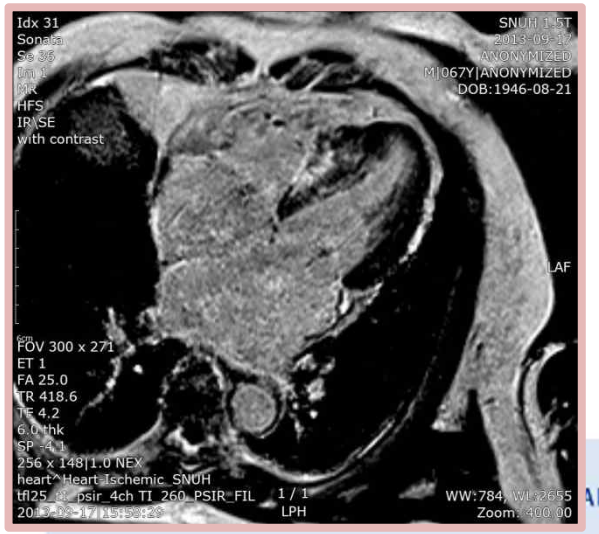
- 69 year-old female

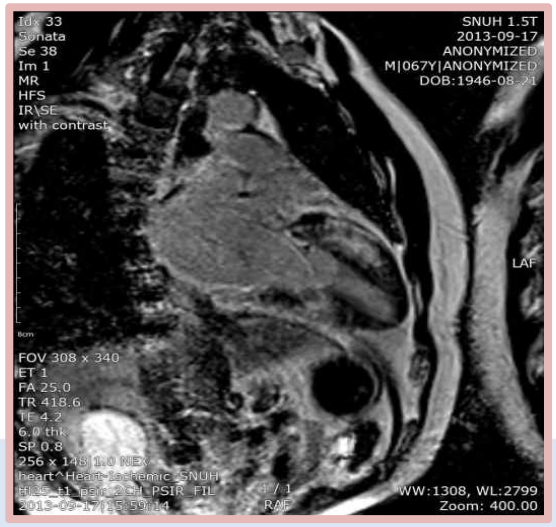


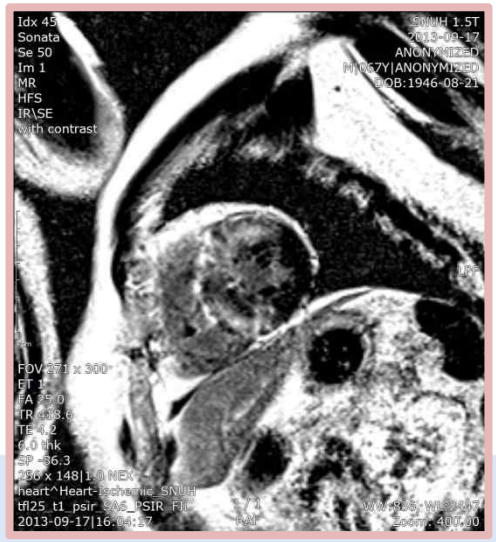




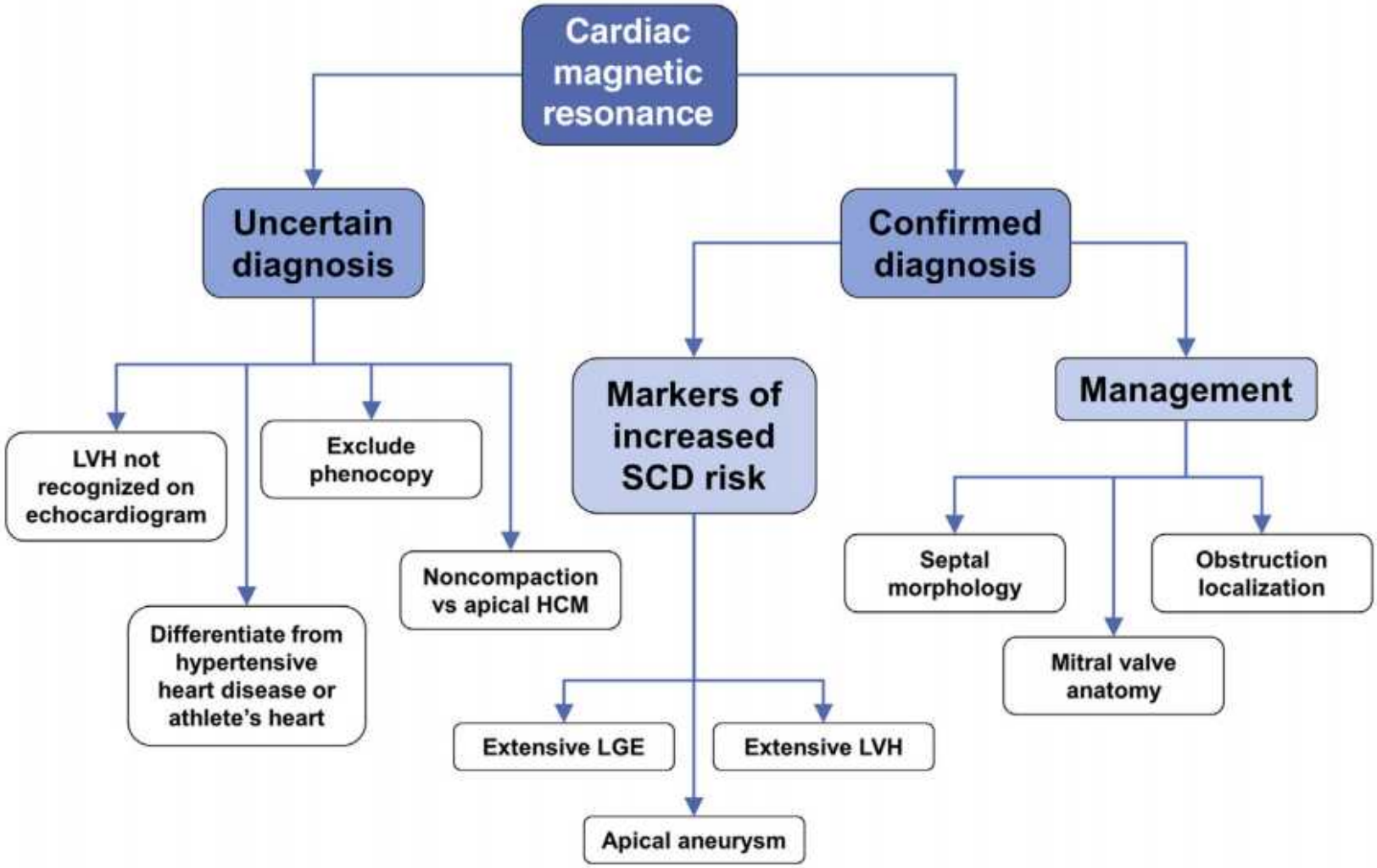
- 66 year-old female







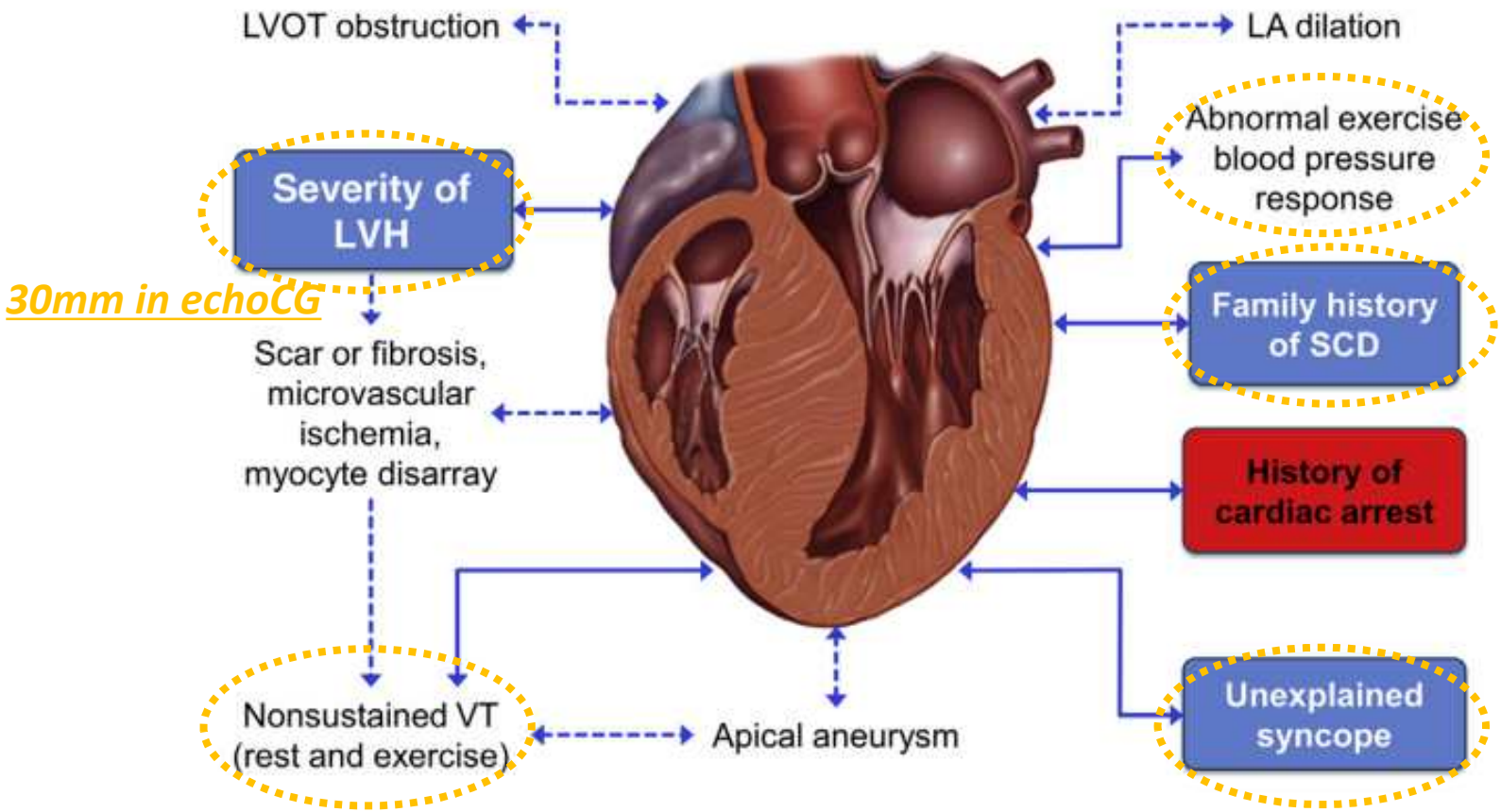
Development of CMR regarding HCMP



Geske JB et al. JACC HF 2018;6:364

Imaging for SCD risk assessment

Conventional risk factors of SCD



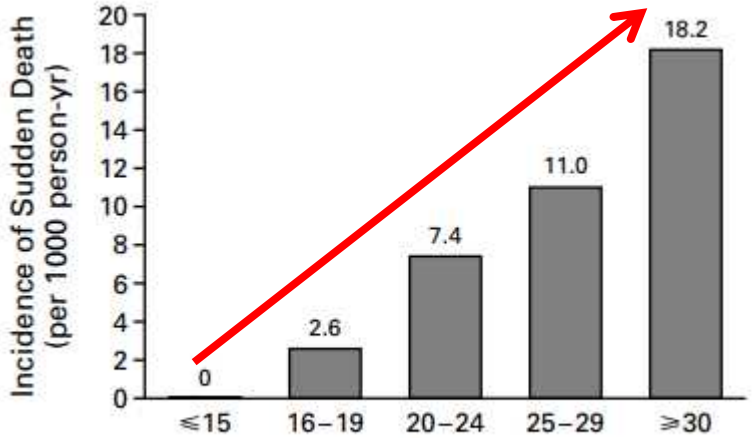
Geske JB et al. JACC HF 2018;6:364

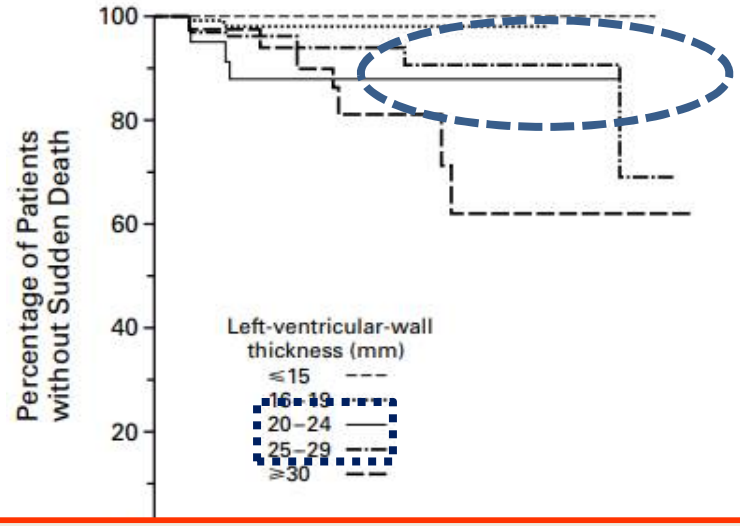
Severity of LVH for prognosis

Table 7 **Major clinical features associated with an increased risk of sudden cardiac death in adults**

Risk Factor	Comment
Age	• The effect of age on SCD has been examined in a number of studies[73,82,99,208,244,372–374] and two have shown a significant association, with an increased risk of SCD in younger patients.[73,99] • Some risk factors appear to be more important in younger patients, most notably, NSVT,[69] severe LVH[375] and unexplained syncope.[99]
Non-sustained ventricular tachycardia	• NSVT (defined as ≥3 consecutive ventricular beats at ≥120 BPM lasting <30 seconds) occurs in 20–30% of patients during ambulatory ECG monitoring and is an independent predictor of SCD.[69,73,83,246,248,374] • There is no evidence that the frequency, duration or rate of NSVT influences the risk of SCD.[69,376]
Maximum left ventricular wall thickness	• The severity and extent of LVH measured by TTE are associated with the risk of SCD.[69,120,121,373] • Several studies have shown the greatest risk of SCD in patients with a maximum wall thickness of ≥30 mm but there are few data in patients with extreme hypertrophy (≥35 mm).[69,73,120,247,248,373,377,378]
Family history of sudden cardiac death at a young age	• While definitions vary,[73,120,372,377] a family history of SCD is usually considered clinically significant when one or more first-degree relatives have died suddenly aged <40 years with or without a diagnosis of HCM, or when SCD has occurred in a first-degree relative at any age with an established diagnosis of HCM.
Syncope	• Syncope is common in patients with HCM but is challenging to assess as it has multiple causes.[379] • Non-neurocardiogenic syncope for which there is no explanation after investigation is associated with increased risk of SCD.[73,83,99,244,246–248] • Episodes within 6 months of evaluation may be more predictive of SCD.[99]
Left atrial diameter	• Two studies have reported a positive association between LA size and SCD.[73,99] There are no data on the association between SCD and LA area and volume. Measurement of LA size is also important in assessing the risk of AF (see section 9.4).
Left ventricular outflow tract obstruction	• A number of studies have reported a significant association with LVOTO and SCD.[73,82,83,246,372,380] Several unanswered questions remain, including the prognostic importance of provocable LVOTO and the impact of treatment (medical or invasive) on SCD.
Exercise blood pressure response	• Approximately one third of adult patients with HCM have an abnormal systolic blood pressure response to exercise characterised by progressive hypotension or a failure to augment the systolic blood pressure that is caused by an inappropriate drop in systemic vascular resistance and a low cardiac output reserve.[241,381] • Various definitions for abnormal blood pressure response in patients with HCM have been reported[69,83,246,377]; for the purposes of this guideline an abnormal blood pressure response is defined as a failure to increase systolic pressure by at least 20 mm Hg from rest to peak exercise or a fall of >20 mm Hg from peak pressure.[237] • Abnormal exercise blood pressure response is associated with a higher risk of SCD in patients aged ≤40 years,[237] but its prognostic significance in patients >40 years of age is unknown.

Severity of LVH for prognosis





dichotomy vs. continuum

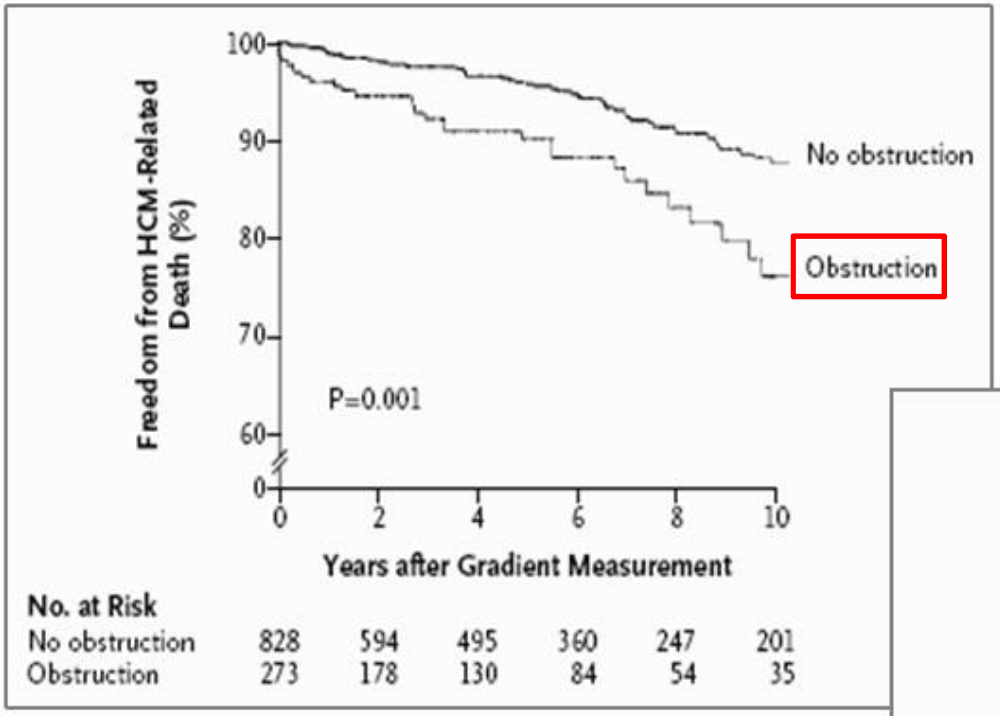
	No risk factors	One risk factor	Two risk factors	Three risk factors	All patients*
Wall thickness					
<15 mm	0·98 (0·96–0·99)	0·96 (0·93–0·98)	0·92 (0·87–0·97)	0·85 (0·73–0·97)	0·96 (0·93–0·98)
15–19 mm	0·97 (0·96–0·99)	0·95 (0·93–0·97)	0·90 (0·85–0·95)	0·81 (0·69–0·93)	0·94 (0·92–0·97)
20–24 mm	0·97 (0·95–0·99)	0·94 (0·91–0·96)	0·88 (0·83–0·92)	0·77 (0·64–0·90)	0·93 (0·90–0·95)
25–29 mm	0·96 (0·93–0·98)	0·92 (0·89–0·95)	0·85 (0·79–0·91)	0·72 (0·57–0·87)	0·91 (0·87–0·94)
≥30mm	0·95 (0·91–0·99)	0·90 (0·85–0·96)	0·81 (0·71–0·91)	0·66 (0·46–0·86)	0·88 (0·82–0·94)
All patients†	0·97 (0·95–0·99)	0·93 (0·91–0·96)	0·87 (0·82–0·92)	0·75 (0·62–0·90)	. .

Number of RF RR 2.0, $p<0.001$ vs. ***LVWTmax*** RR 1.26, $p=0.058$

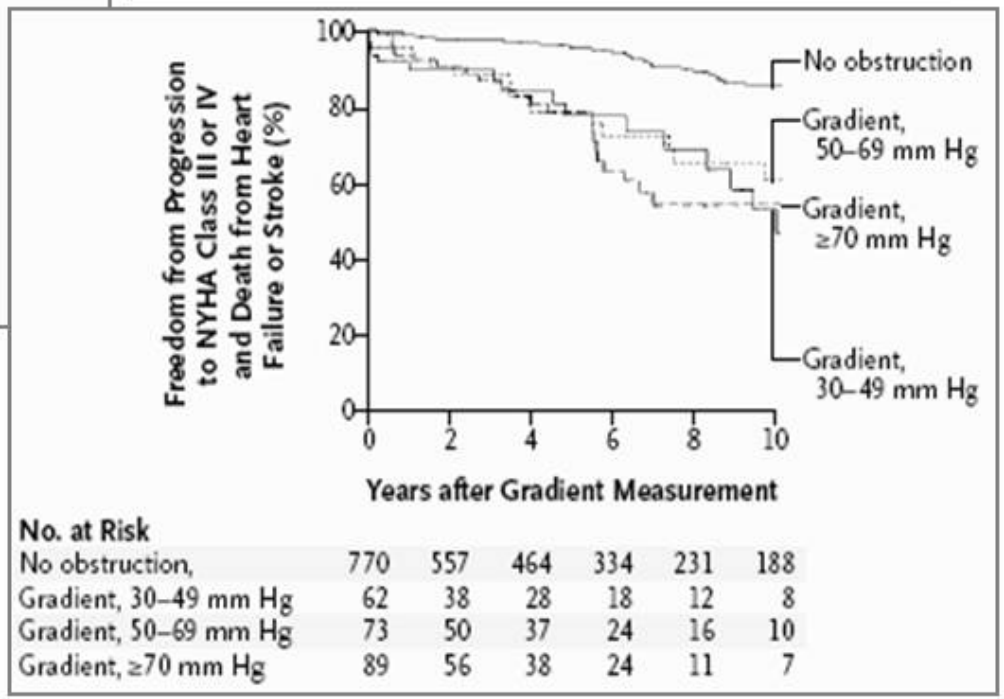
Spirito P et al. NEJM 2000;342:1778
Elliott PM et al. Lancet 2001;357:420

LVOT obstruction

1,101 HCM patients

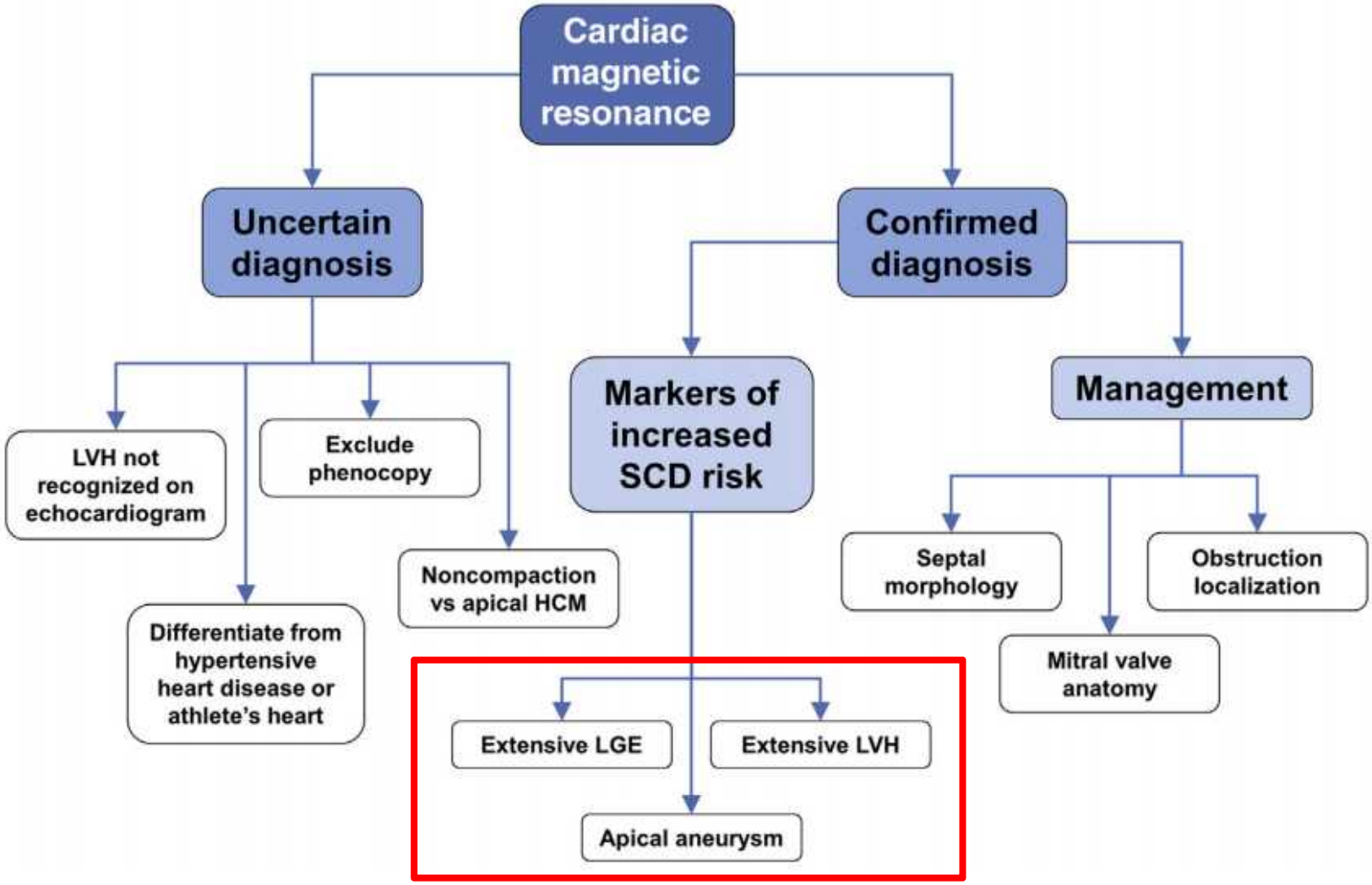


Variable	Death from Any Cause	
	Relative Risk (95% CI)	P Value
Left ventricular outflow obstruction (≥30 mm Hg)	1.6 (1.1–2.2)	0.02
NYHA class II, III, or IV at entry	1.5 (1.1–2.1)	0.02
Paroxysmal or chronic atrial fibrillation	1.4 (1.0–1.9)	0.04
Maximal left ventricular thickness ≥30 mm	1.6 (1.1–2.4)	0.01



Geske JB et al. JACC HF 2018;6:364

Development of CMR regarding HCMP



Geske JB et al. JACC HF 2018;6:364

Relation to occurrence of frequent of arrhythmias

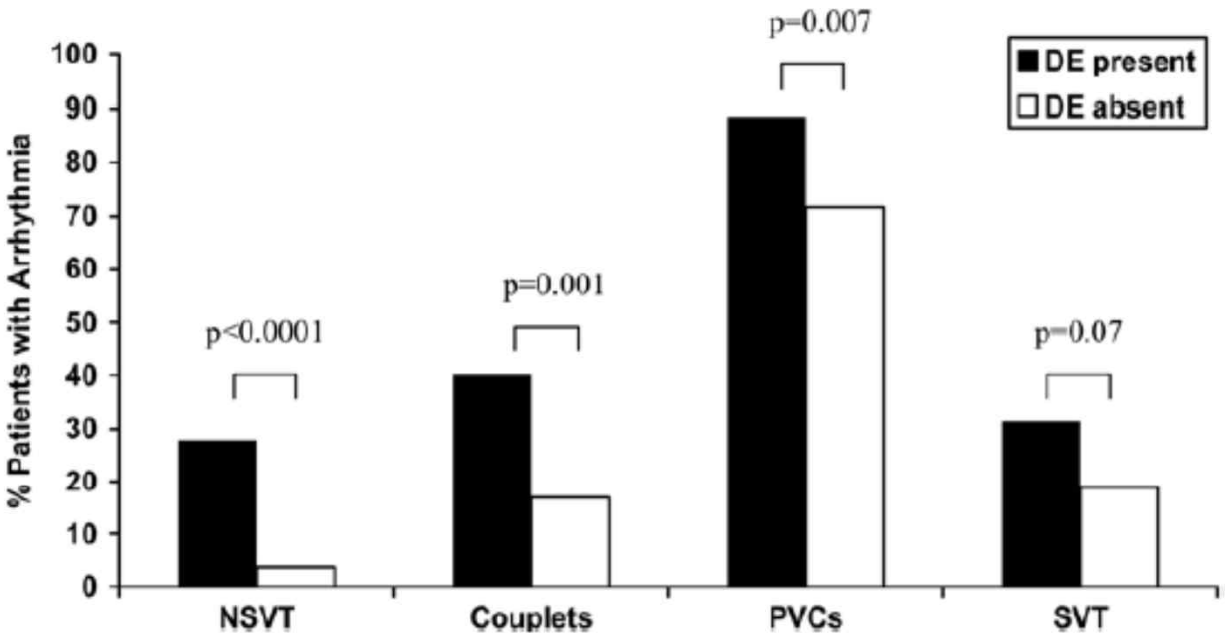


Figure 1 **Prevalence of Arrhythmias on 24-h Holter ECG With Respect to DE in 177 HCM Patients**

DE = delayed enhancement; ECG = electrocardiogram; HCM = hypertrophic cardiomyopathy; NSVT = nonsustained ventricular tachycardia; PVC = premature ventricular contraction; SVT = supraventricular tachycardia.

Adabag AS et al. JACC 2008;51:1369

Relation to VT/VF and risk of sudden death

Table 4 Logistic regression analysis for clinical arrhythmias

Variable	OR	95% CI for OR	*P*-value
Univariate analysis			
DCE score	1.080	1.030–1.131	0.001
LVMI	1.010	1.000–1.021	0.05
Age	1.015	0.987–1.043	0.30
Male gender	1.184	0.209–1.605	0.29
DCE present	0.400	0.117–1.366	0.14
MaxLVWT	1.047	0.989–1.109	0.11
MEDT	1.118	0.977–1.278	0.10
LVEDVI	1.008	0.988–1.028	0.42
LVEF	0.968	0.929–1.008	0.12
Multivariable analysis			
DCE score	1.073	1.023–1.125	0.004
LVMI	1.006	0.996–1.016	0.22

Table 6 Logistic regression analysis for sudden cardiac death (SCD) risk (Group LR vs. Group R)

Variable	OR	95% CI for OR	*P*-value
Univariate analysis			
MaxLVWT	1.201	1.096–1.315	<0.0001
LVMI	1.018	1.005–1.032	0.007
MEDT	1.349	1.128–1.615	0.001
DCE score	1.089	1.035–1.145	0.001
Age	0.991	0.967–1.016	0.50
Male gender	1.184	0.490–2.860	0.71
DCE present	0.400	0.117–1.366	0.14
LVEDVI	1.005	0.986–1.024	0.62
LVEF	0.980	0.942–1.019	0.30
Multivariable analysis			
MaxLVWT	1.181	1.054–1.324	0.004
DCE score	1.018	1.003–1.034	0.019
LVMI	1.001	0.979–1.024	0.916
MEDT	1.016	0.727–1.419	0.927

Clinical arrhythmia: cardiac arrest, sustained VT, and non-sustained VT

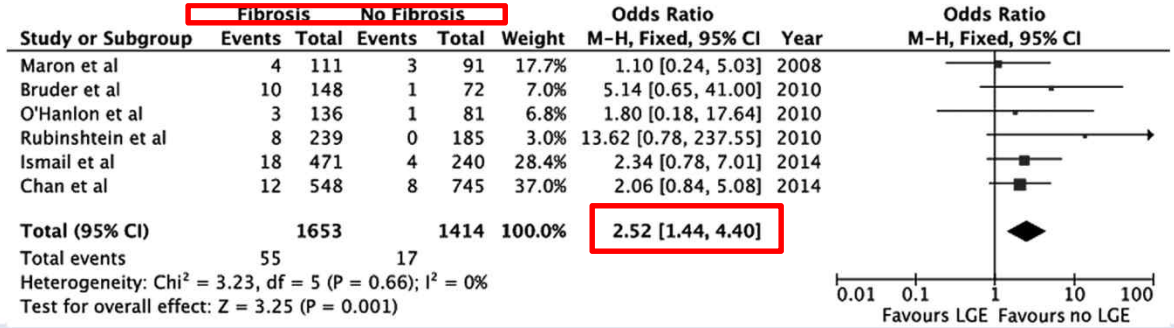
Leonardi S et al. EHJ 2009;30:2003

Link between LGE and SCD

Table 1 Interventions and characteristics of individual studies

Study	Study design	Sample size: No fibrosis	Sample size: Fibrosis	Sample size: Whole cohort	Mean age (years)	Extent of LGE (%)	Exclusion criteria	Primary endpoints	Secondary endpoints	Follow-up (years)
Ismail *et al* (2014)[6]	Prospective observational study	240	471	711	56	9.5	Prior myectomy, alcohol septal ablation, previous MI, contraindications to CMR and gadolinium-based contrast agents	SCD or aborted SCD	Composite of cardiovascular mortality, aborted SCD or cardiac transplantation and all-cause mortality	3.5
Chan *et al* (2014)[4]	Prospective observational study	745	548	1293	46	9	Prior ICD, VT/VF, known CAD or MI, septal myectomy or alcohol ablation	SCD or aborted SCD	All-cause mortality, ICD shock, HF death, heart transplantation, progression of HF, non-cardiac death	3.3
O'Hanlon *et al* (2010)[8]	Prospective observational study	81	136	217	51	15.5	Patients with known CAD or MI, septal myectomy or alcohol ablation	Composite of cardiovascular death, hospital stay, VT/VF or appropriate ICD shock	Composite of unplanned HF hospital stay, progression of HF or HF-related death Composite of sustained VT/VF, appropriate ICD discharge or SCD	3.1
Bruder *et al* (2010)[5]	Prospective observational study	72	148	220	57	3.2	Known CAD, aortic stenosis, amyloidosis, hypertension, contraindications to CMR, prior septal myectomy or alcohol ablation	All-cause death and cardiac death (including SCD, HF and aborted SCD)		3
Rubinshtein *et al* (2010)	Retrospective analysis	185	239	424	55	6.2	Prior myectomy, alcohol septal ablation, previous MI, contraindications to CMR and gadolinium-based contrast agents	SCD or appropriate ICD therapy	–	3.6
Maron *et al* (2008)[7]	Prospective observational study	91	111	202	42	9	Prior myectomy, alcohol septal ablation, previous MI or obstructive, contraindications to CMR and gadolinium-based contrast agents	Composite of SCD, appropriate ICD shock and progressive HF symptoms	–	1.8

CAD, coronary artery disease; CMR, cardiac MRI; HF, heart failure; ICD, implantable cardioverter defibrillator; LGE, late gadolinium enhancement; MI, myocardial infarction; SCD, sudden cardiac death; VF, ventricular fibrillation; VT, ventricular tachycardia.



OR 2.52
95% CI 1.44 to 4.4
p=0.001

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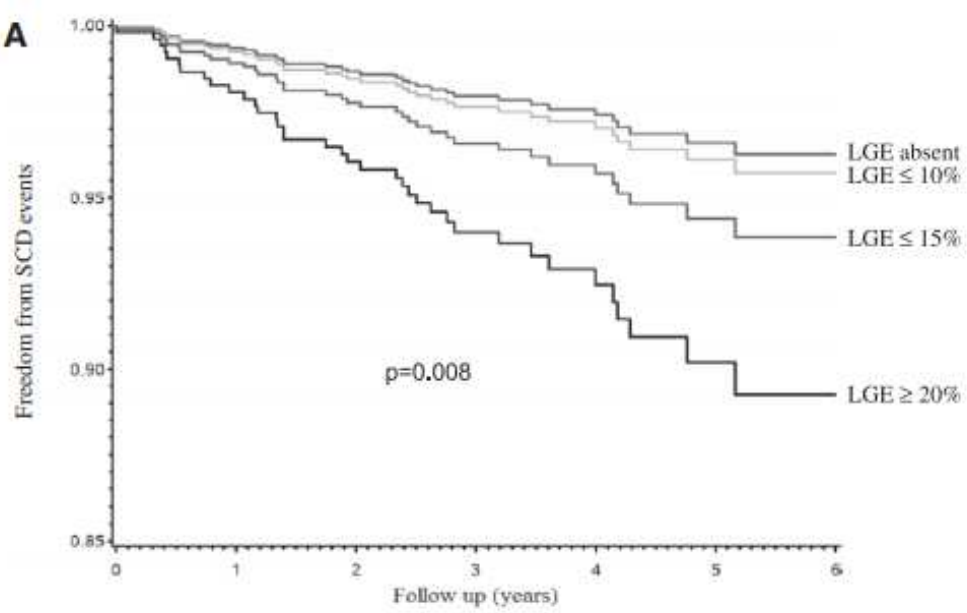
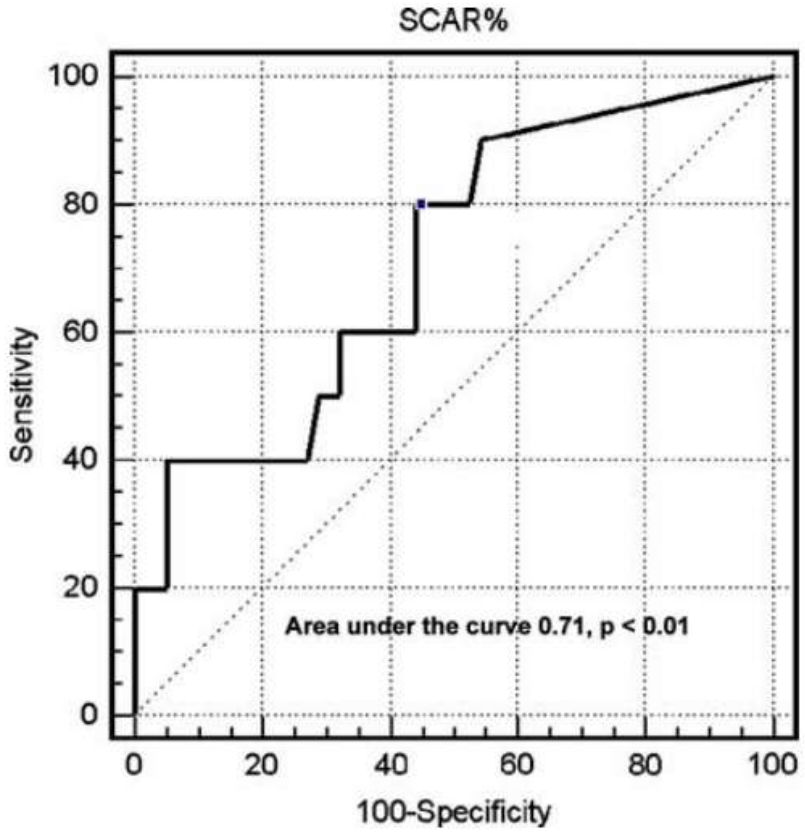
Briasoulis A, et al. Heart 2015;101:1406

Prognostic value of LGE in HCMP

Extent of LGE



1,293 HCM patients, followed up 3.3 yrs → 37 SCD (3%)



68 HCM patients, monitoring for 39hrs → 9 NSVT (13%)

Kwon DH et al. Int J Cardiovasc Img 2008;24:617
Chan RH et al. Circulation 2014;130:484

Prognostic value of LGE in HCMP

Presence of LGE

A) Cardiac Death

Study	Odds Ratio(95% CI)	p-value
Bruder	8.00 (1.04-61.87)	0.046
O'Hanlon	5.00 (0.61-40.73)	0.133
Maron	0.54 (0.08-3.29)	0.503
Rubinshtein	10.33 (0.58-184.51)	0.112
Pooled	2.92 (1.01-8.42)	0.047

B) SCD/Aborted SCD

Study	Odds Ratio(95% CI)	p-value
Bruder	5.15 (0.65-41.00)	0.112
O'Hanlon	1.81 (0.19-17.64)	0.612
Maron	1.10 (0.24-5.03)	0.906
Rubinshtein	13.62 (0.78-237.55)	0.073
Pooled	2.39 (0.87-6.58)	0.091

C) HF Death

Study	Odds Ratio(95% CI)	p-value
Bruder	5.56 (0.30-101.90)	0.248
O'Hanlon	8.12 (0.45-146.04)	0.115
Rubinshtein	3.91 (0.19-81.83)	0.380
Pooled	5.68 (1.04-31.07)	0.045

D) All Cause Mortality

Study	Odds Ratio(95% CI)	p-value
Bruder	5.47 (1.24-24.08)	0.025
Rubinshtein	3.58 (0.76-16.78)	0.106
Pooled	4.46 (1.53-13.01)	0.006

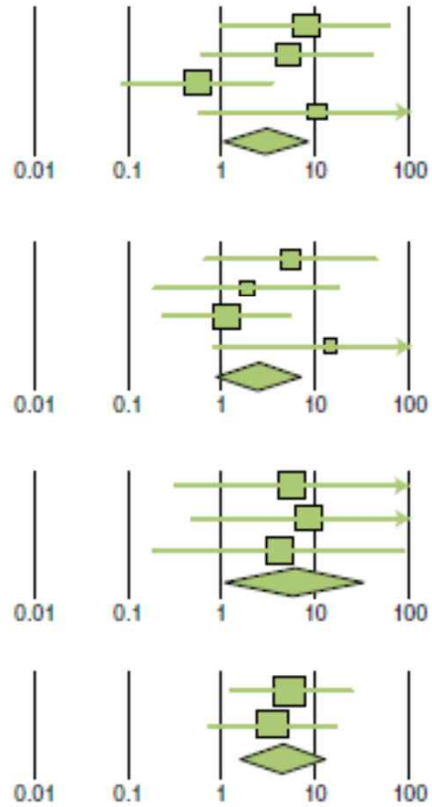


Figure 2. Forrest Plots and Pooled Odds Ratios for Clinical Endpoints

The presence of late gadolinium enhancement by cardiac magnetic resonance predicted (**A**) cardiac death, (**C**) heart failure (HF) death, and (**D**) all-cause mortality. Additionally, there was a trend toward significance for (**B**) prediction of sudden cardiac death (SCD)/aborted SCD. CI = confidence interval.

Green JJ et al. JACC CVI 2012;5:370

Prediction of major adverse events

	No Cardiac Mortality (n = 204)	Cardiac Mortality (n = 16)	p Value	OR (95% CI)
Age, yrs	57.0 (46.0–68.0)	61.5 (57.0–73.0)	<0.05	1.04 (1.00–1.08)
Pattern				
Septal	84.3 (172)	81.2 (13)	0.72	0.81 (0.22–2.99)
Apical	7.4 (15)	12.5 (2)	0.36	1.80 (0.37–8.67)
Concentric	8.3 (17)	6.2 (1)	1.00	0.73 (0.09–5.90)
CMR parameter				
LVEF, %	71.0 (64.8–76.9)	68.0 (51.2–75.2)	<0.05	0.95 (0.92–0.99)
Maximal wall thickness, mm	19.0 (16.0–22.5)	20.0 (17.5–24.5)	0.35	1.05 (0.95–1.16)
LV mass, g	154.8 (126.8–190.9)	186.0 (150.3–229.7)	0.05	1.01 (1.00–1.01)
LV mass index, g/m^2	81.4 (66.2–95.3)	97.1 (82.0–126.0)	<0.01	1.02 (1.00–1.03)
LVOT obstruction, %	30.9 (63.0)	37.5 (6.0)	0.58	1.34 (0.47–3.86)
LGE	65.2 (133.0)	93.8 (15.0)	<0.05	8.01 (1.04–61.9)
LGE, g	1.8 (0.0–7.4)	15.6 (5.9–23.4)	<0.001	1.02 (1.00–1.04)
LGE, % LV	1.1 (0.0–4.6)	7.4 (3.4–17.3)	<0.001	1.05 (1.01–1.09)
Surface area LGE, mm^2	75.6 (0.0–272.3)	393.1 (185.7–849.9)	<0.001	1.00 (1.00–1.00)
Surface area/LV mass, mm^2/g	0.5 (0.0–1.7)	2.0 (1.3–4.8)	<0.001	1.20 (1.05–1.38)
SCD risk factors				
Maximal wall thickness >30 mm	3.4 (7.0)	6.3 (1.0)	0.46	1.88 (0.22–16.27)
History of spontaneous VT	4.9 (10.0)	12.5 (2.0)	0.21	2.77 (0.55–13.90)
Family history of SCD	4.4 (9.0)	6.3 (1.0)	0.54	1.44 (0.17–12.18)
Unexplained syncope	4.9 (10.0)	12.5 (2.0)	0.21	2.77 (0.55–13.90)
LVOT obstruction >30 mm Hg	12.2 (22.0)	13.3 (2.0)	1.00	1.10 (0.23–5.23)
Number of SCD risk factors				
0	76.5 (156.0)	68.8 (11.0)	0.54	0.68 (0.22–2.04)
1	19.6 (40.0)	18.8 (3.0)	1.00	0.95 (0.26–3.48)
2	2.9 (6.0)	6.3 (1.0)	0.38	2.20 (0.25–19.48)

220 pts, 22 pts events, FU 1090 days, LGE 67%

O'Hanlon R et al. JACC 2010;56:867

Prediction of major adverse events

Fibrosis and development of primary end point and annual probability of primary end point

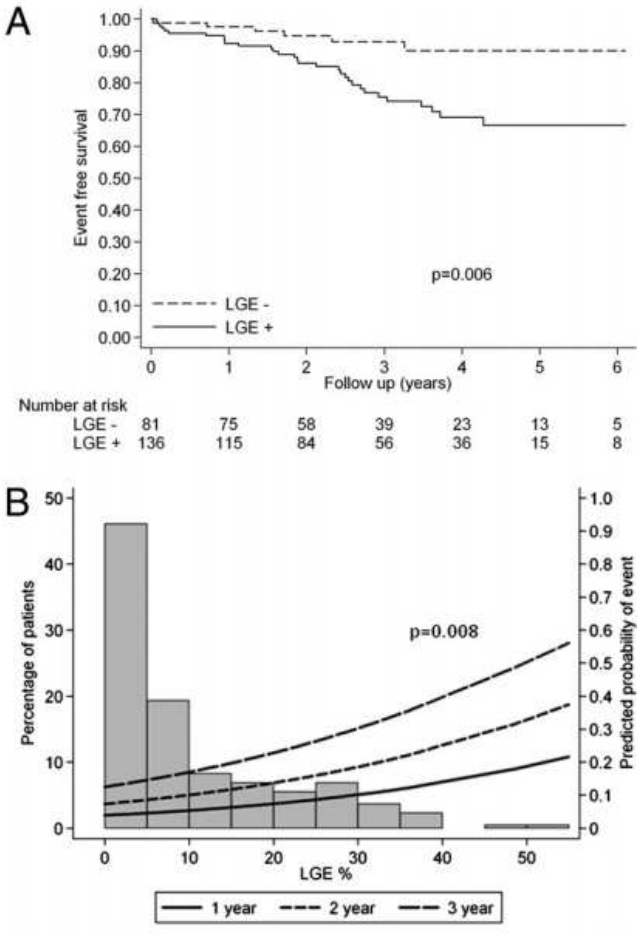


Table 3 Breakdown of Events Contributing to Overall MACE Rate in 217 HCM Patients With and Without Fibrosis

Outcome	No Fibrosis (n = 81)	Fibrosis (n = 136)	Total (n = 217)	HR	95% CI	p Value
Primary outcome	6 (7.4)	34 (25.0)	40 (18.4)	3.367	1.406–8.063	0.006
CV mortality	1 (1.2)	8 (5.9)	9 (4.2)	4.452	0.548–36.204	0.163
Unplanned CV hospital stay	5 (6.2)	24 (17.7)	29 (13.4)	2.825	1.072–7.448	0.036
VT/VF	1 (1.2)	8 (5.9)	9 (4.2)	4.973	0.622–39.762	0.131
ICD discharge	0 (0)	2 (1.5)	2 (0.9)	NA	—	—
Sudden death	1 (1.2)	1 (0.7)	2 (0.9)	0.648	0.041–10.360	0.759
HF death	0 (0)	6 (4.4)	6 (2.8)	—	—	—
CVA death	0 (0)	1 (0.7)	1 (0.4)	NA	—	—

CI = confidence interval; CV = cardiovascular; CVA = cardiovascular accident; HR = hazard ratio; ICD = implantable cardioverter-defibrillator; VT/VF = ventricular tachycardia/ventricular fibrillation.

O'Hanlon R et al. JACC 2010;56:867

LGE in HCMP

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- Substrate for **ventricular arrhythmia**
- Associated with **ventricular remodeling and heart failure**
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220 pts, 22 pts events, FU 1090 days, LGE 67%

O'Hanlon R et al. JACC 2010;56:867

Link between LGE and SCD

Meta-regression analysis

We performed a meta-regression analysis, which demonstrated that the extent of LGE (%) was not significantly related to SCD/aborted-SCD risk (p=0.35), all-cause mortality (p=0.084), cardiac death (p=0.59) and HF death (p=0.99) (see online supplementary table).

Table 1 Interventions and characteristics of individual studies

Study	Study design	Sample size: No fibrosis	Sample size: Fibrosis	Whole cohort	Mean age (years)	Extent of LGE (%)				w-up s)
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O'Hanlon *et al* (2010)[8]	Prospective observational study	81	136 **63%**	217	51	15.5	Patients with known CAD or MI, septal myectomy or alcohol ablation	Composite of cardiovascular death, hospital stay, VT/VF or appropriate ICD shock	Composite of unplanned HF hospital stay, progression of HF or HF-related death Composite of sustained VT/VF, appropriate ICD discharge or SCD	3.1
Bruder *et al* (2010)[5]	Prospective observational study	72	148 **67%**	220	57	3.2	Known CAD, aortic stenosis, amyloidosis, hypertension, contraindications to CMR, prior septal myectomy or alcohol ablation	All-cause death and cardiac death (including SCD, HF and aborted SCD)		3
Rubinshtein *et al* (2010)	Retrospective analysis	185	239 **56%**	424	55	6.2	Prior myectomy, alcohol septal ablation, previous MI, contraindications to CMR and gadolinium-based contrast agents	SCD or appropriate ICD therapy	–	3.6
Maron *et al* (2008)[7]	Prospective observational study	91	111 **55%**	202	42	9	Prior myectomy, alcohol septal ablation, previous MI or obstructive, contraindications to CMR and gadolinium-based contrast agents	Composite of SCD, appropriate ICD shock and progressive HF symptoms	–	1.8

CAD, coronary artery disease; CMR, cardiac MRI; HF, heart failure; ICD, implantable cardioverter defibrillator; LGE, late gadolinium enhancement; MI, myocardial infarction; SCD, sudden cardiac death; VF, ventricular fibrillation; VT, ventricular tachycardia.

The extent of LGE was not significantly related to the risk of SCD.

Briasoulis A, et al. Heart 2015;101:1406

LGE in HCMP

- Correlates with conventional **risk factors of sudden death**
- Substrate for **ventricular arrhythmia**
- Associated with **ventricular remodeling and heart failure**
- Associated with **an increased risk of major adverse events**
- The risk is proportional with increased **amounts of LGE-detected fibrosis**

- **Limitations**
  - **Presence of LGE: too common**
  - **LGE%: cut-off value, standardization, reproducibility issue**
  - **Not a 1:1 relationship between the presence of LGE and cardiac death**
- **Combination prediction model**
- **Conventional low risk group**
- **Large longitudinal follow-up multicenter studies are needed.**

LGE in HCMP

5.5.3 Late Gadolinium Enhancement and Prognosis

The association between LGE and long-term outcomes has been examined in six studies,[138–143] four of which are included in a meta-analysis (*Web Table 4*).[144] All published studies are limited by selection and referral bias, incomplete risk assessment and differences in scanning protocols and LGE quantification. The pooled data support a relationship between LGE and cardiovascular mortality, heart failure death and all-cause death, but show only a trend towards an increased risk of SCD.[144] Late gadolinium enhancement is associated with NSVT on Holter monitoring.[140,142]

On balance, the extent of LGE on CMR has some utility in predicting cardiovascular mortality, but current data do not support the use of LGE in prediction of SCD risk.

Summaries

- **Echocardiography** is central to the diagnosis and monitoring of HCMP.
  - Assessment of LV wall thickness
  - Associated abnormalities of the mitral valve and LVOT
  - Left atrial enlargement, diastolic function
  - Systolic function
  - Differential diagnosis
- **Cardiovascular magnetic resonance imaging** of patients with known or suspected HCM should be in line with current guideline recommendation and should be performed and interpreted by teams experienced in cardiac imaging and in the evaluation of heart muscle disease
  - Assessment of ventricular morphology and function
  - Myocardial fibrosis (late gadolinium enhancement, LGE)
  - LGE and prognosis
  - Differential diagnosis

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Ji-Hyun Lee, MD, PhD

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Sejong General Hospital, Bucheon Arrhythmia Clinic
Won-Seok Choi, MD

The Catholic University of Korea
Kyung-Do Han, PhD
Jin-Hyung Jung, BSc

Thank you for your attention

SNUH 서울대학교병원
SEOUL NATIONAL UNIVERSITY HOSPITAL

Apical aneurysm

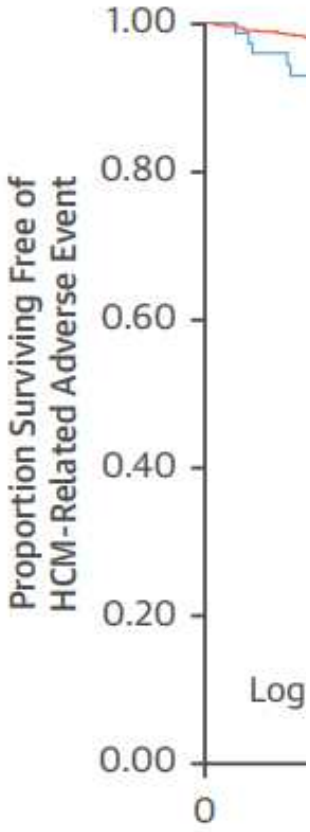
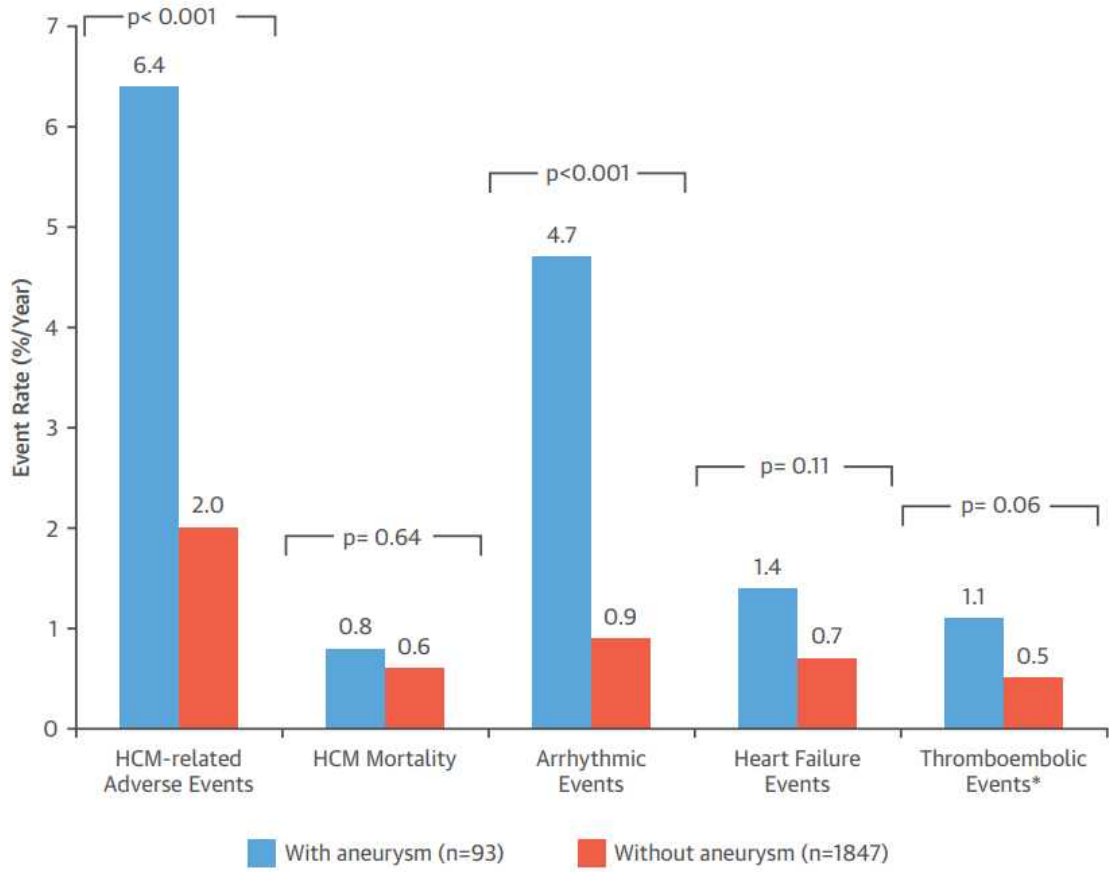


FIGURE 4 Clinical Outcome in 93 Patients With LV Apical Aneurysm Compared With HCM Cohort Without Aneurysm (n = 1,847)



Rowin EJ et al. JACC 2017;69:761