Alfredo PIZZUTI

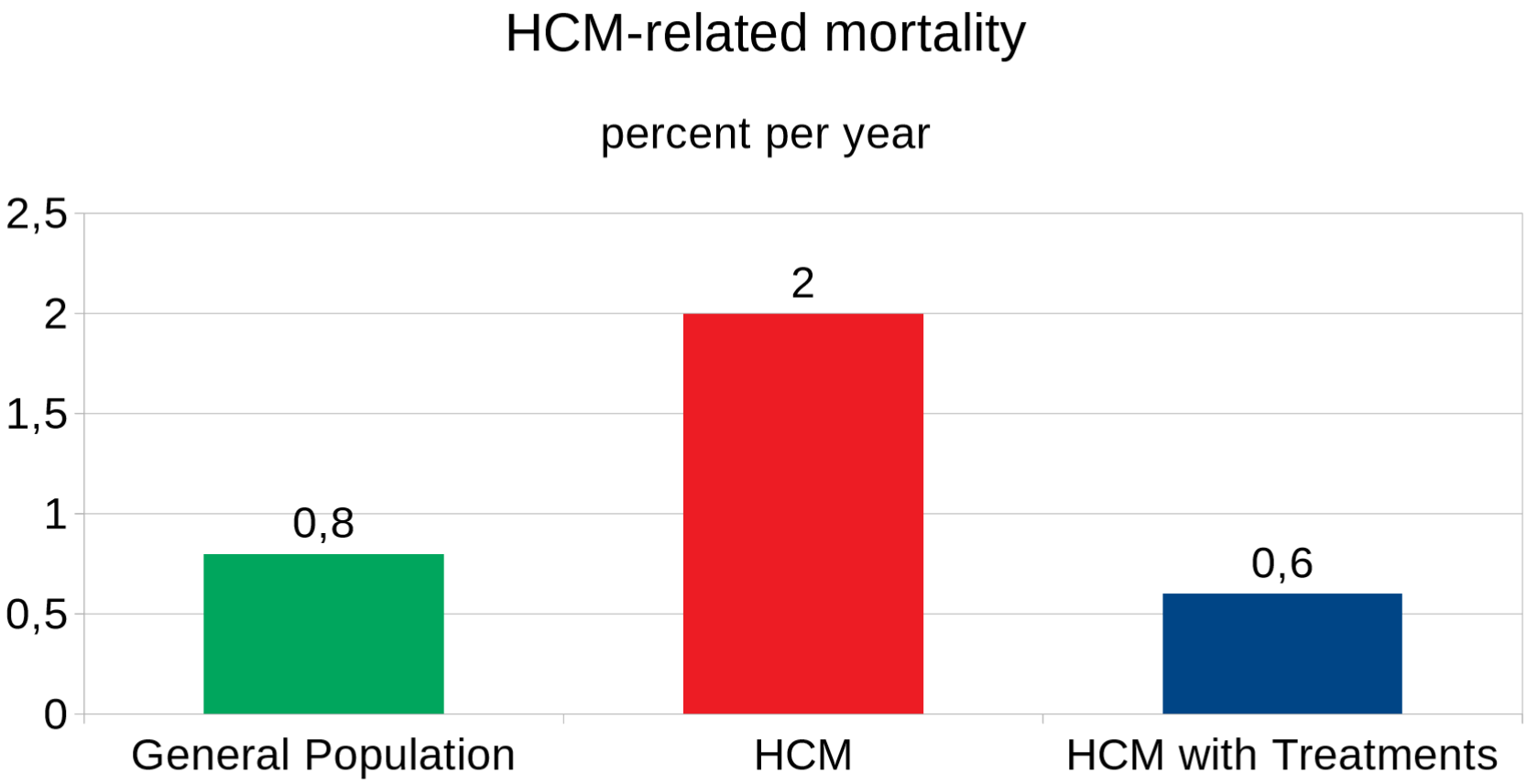
SC Cardiologia
AO Ordine Mauriziano
Torino

TURIN
October
24th-26th
2019

31 GIORNATE CARDIOLOGICHE TORINESI

Everything you always wanted to know about Hypertrophic Cardiomyopathy

Hypertrophic Cardiomyopathy



Maron BJ, N Engl J Med 2018; 379:655

Risk factors for Sudden Death

- **Age**
- **Family history of Sudden Death (SD)**
- **Unexplained Syncope**
- **Left Ventricular wall thickness**
- **Left Ventricular outflow gradient**
- **Non Sustained Ventricular Tachycardia**

Hypertrophic Cardiomyopathy

Risk of Sudden Death and Outcome in Patients With Hypertrophic Cardiomyopathy With Benign Presentation and Without Risk Factors

Paolo Spirito, MD[a,*], Camillo Autore, MD[b], Francesco Formisano, MD[a], Gabriele Egidy Assenza, MD[b], Elena Biagini, MD[c], Tammy S. Haas, RN[d], Sergio Bongioanni, MD[e], Christopher Semsarian, MD[f], Emmanuela Devoto, MD[a], Beatrice Musumeci, MD[b], Francesco Lai, MD[c], Laura Yeates, BSc[f], Maria Rosa Conte, MD[e], Claudio Rapezzi, MD[c], Luca Boni, MD[g], and Barry J. Maron, MD[d]

- **absence of each of the conventional major sudden death risk factors**
- NYHA I or II
- No Atrial Fibrillation
- No myectomy, septal ablation or ICD implant
- LVEF >50%
- Age ≥10 and ≤75 years

N = 653; f-u 5.3 y

SD: 0.6% per year

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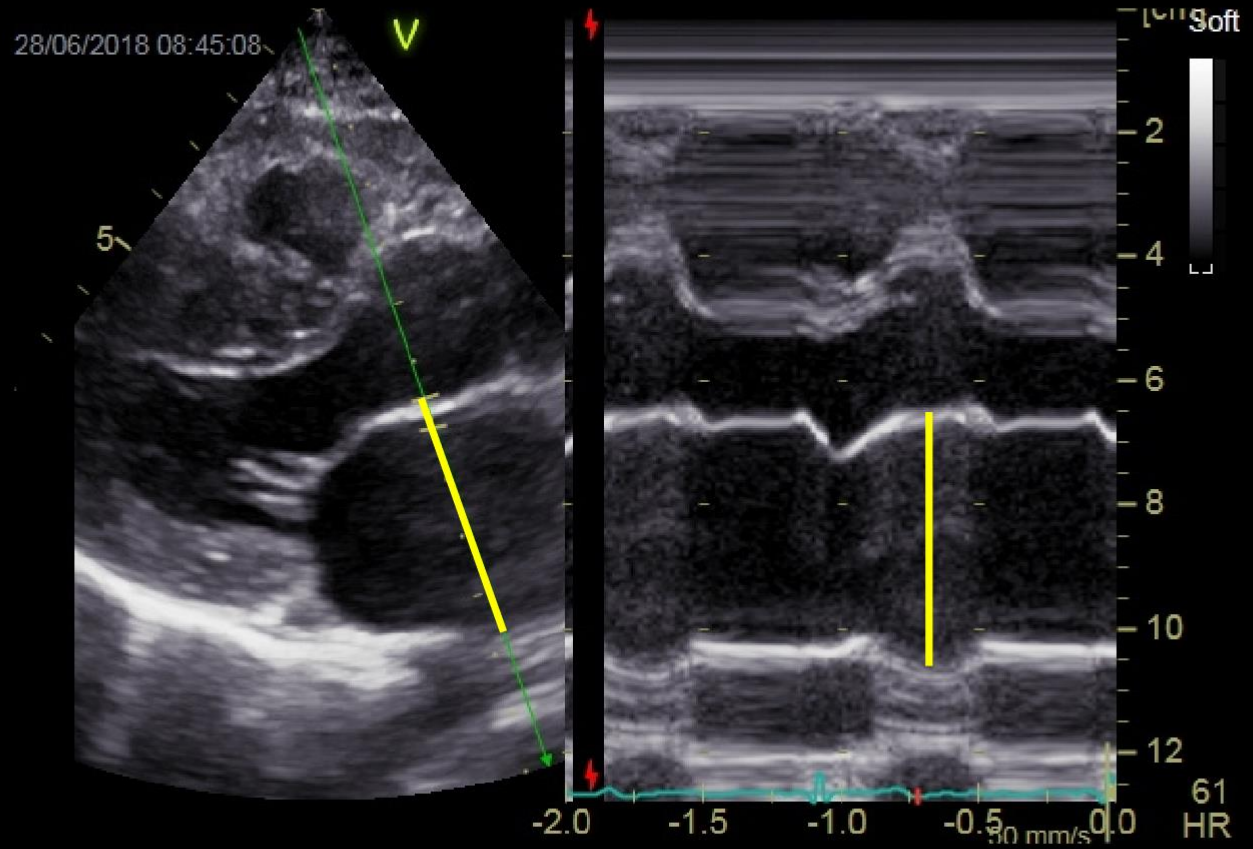
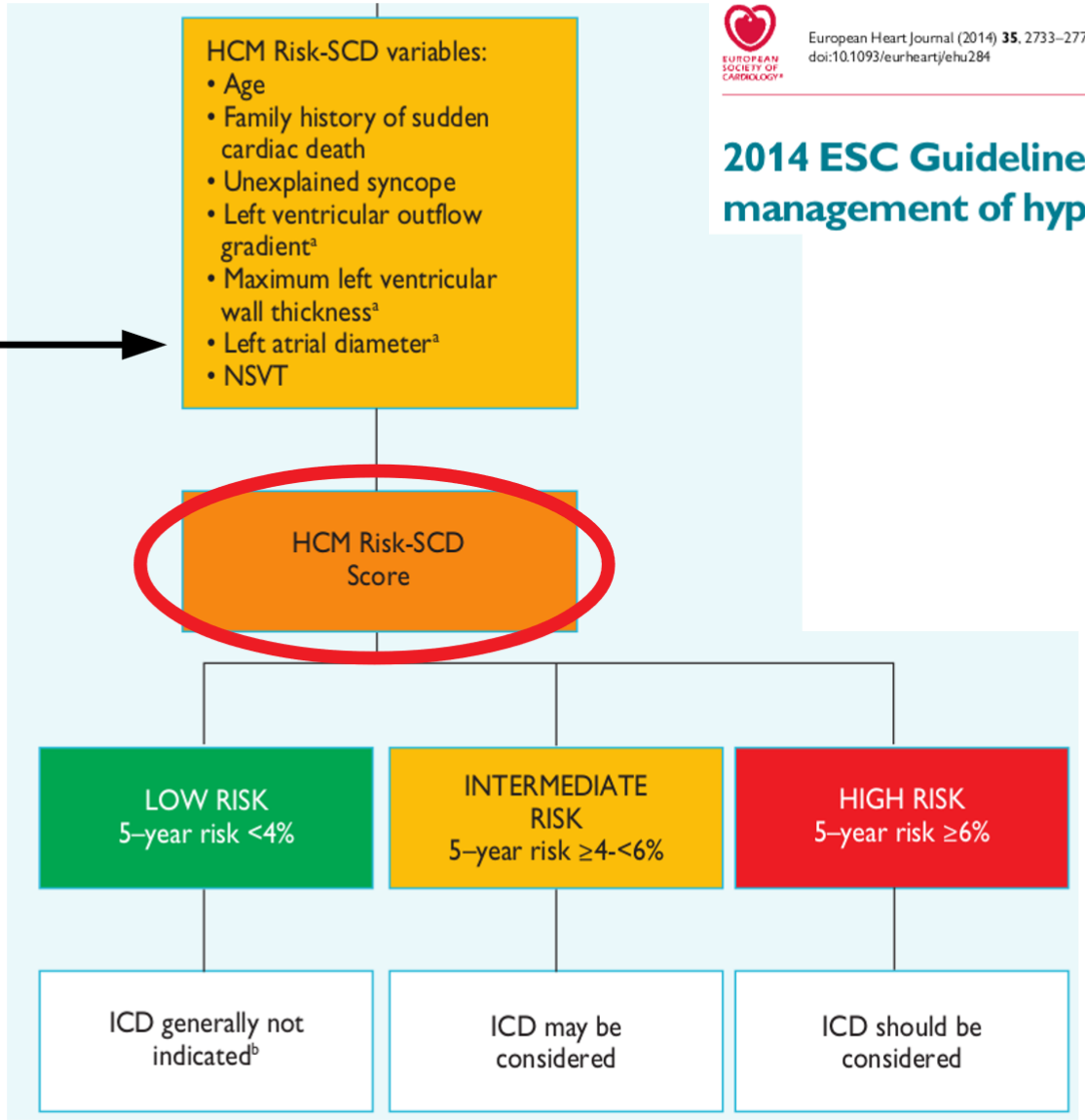
	Relative Risk (95% IC)	*p*
Age	0.97 (0.94–0.99)	**0.020**
Sex	-	0.43
Left Ventricular Outflow Obstruction	-	0.822
Wall thickness	-	0.646
Left Atrial diameter (mm):		
< 41	1	**0.009**
41 - 50	3.11 (1.13-8.54)	
> 50	8.01 (2.08-30.9)	

Hypertrophic Cardiomyopathy



Hypertrophic Cardiomyopathy



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

ESC GUIDELINES

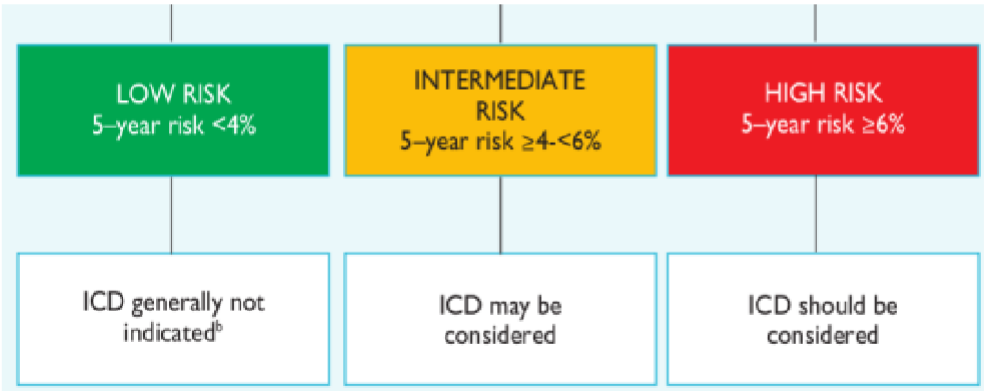
2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy

Hypertrophic Cardiomyopathy

Probability of Sudden Death at 5 years

$$P=1 - 0.998^{\exp(\text{Prognostic Index})}$$

$$\begin{aligned}
& 0.1593 * \text{Maximal Wall Thickness (mm)} \\
- \; & 0.0029 * \text{MWT}^2 \ (\text{mm}^2) \\
+ \; & 0.0259 * \text{Left Atrial Dimension (mm)} \\
+ \; & 0.0044 * \text{Outflow gradient (mmHg)} \\
+ \; & 0.4583 * \text{Family History of SD} \\
+ \; & 0.8263 * \text{NSVT} \\
+ \; & 0.7165 * \text{Syncope} \\
- \; & 0.0179 * \text{Age (years)} \\
\hline
= \; & \text{Prognostic Index}
\end{aligned}$$



Hypertrophic Cardiomyopathy

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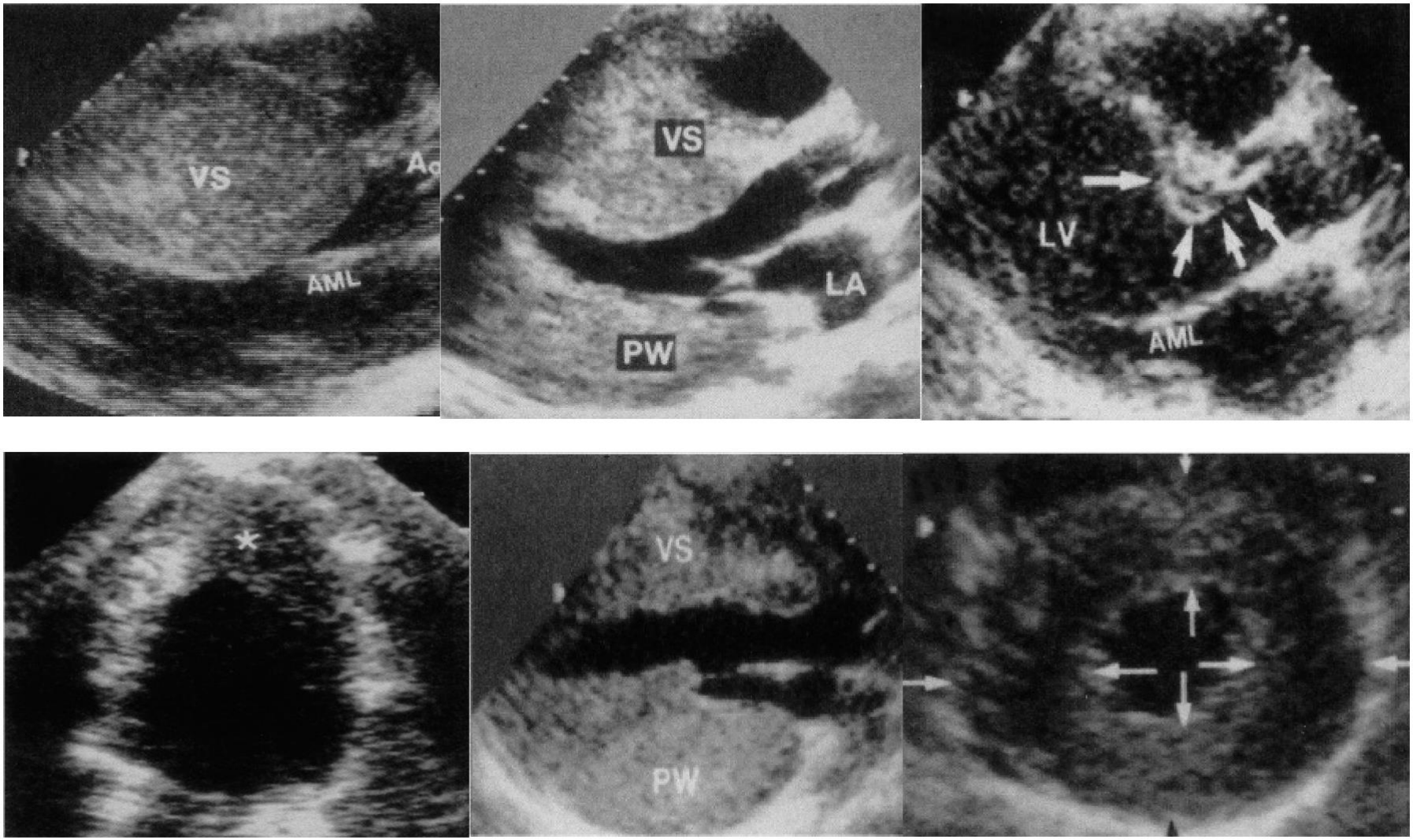
=\ &\text{Prognostic Index}
\end{aligned}$$

*However, when applied to individual patients with HCM, the risk score has **low sensitivity** for making clinically relevant decisions about implantable cardioverter–defibrillator (ICD) placement, significantly underidentifying high-risk patients*

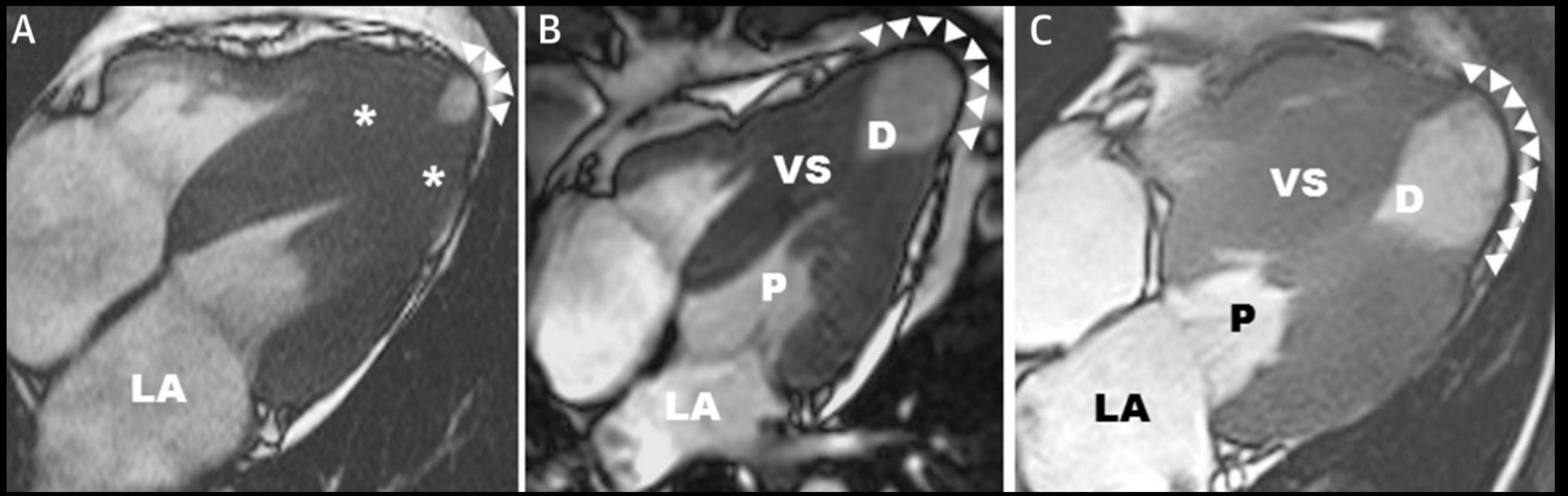
Maron BJ, N Engl J Med 2018; 379:655

Hypertrophic Cardiomyopathy



Klues HG, JACC 1995;26:1699

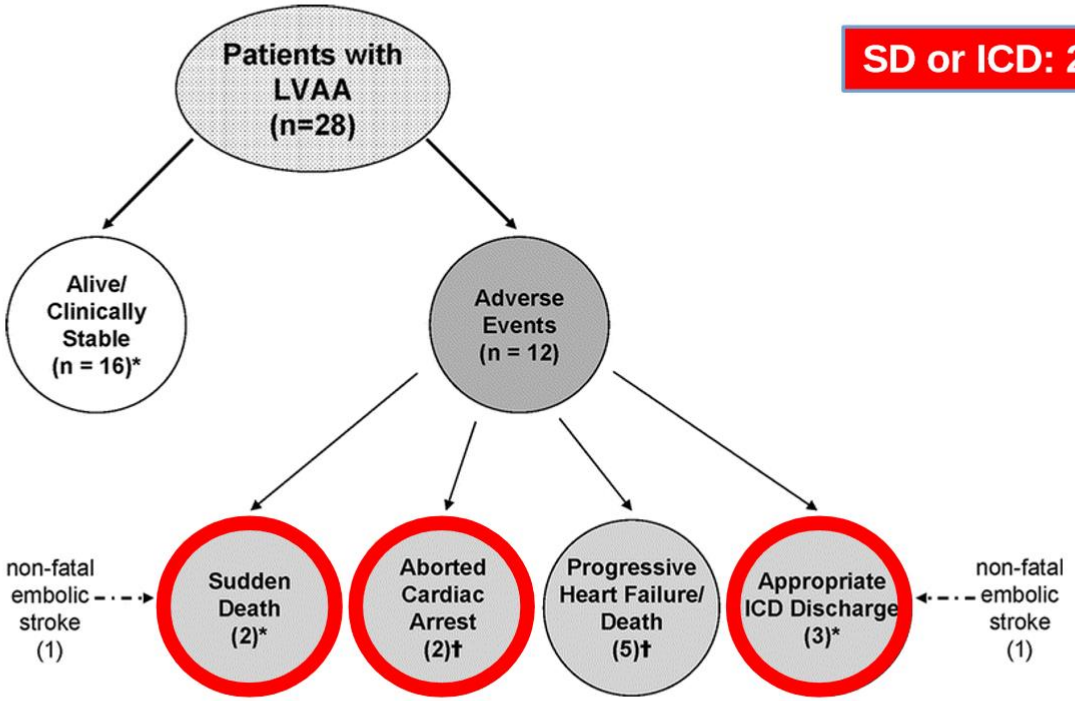
Apical Aneurysm in Hypertrophic Cardiomyopathy



Hypertrophic Cardiomyopathy

Prevalence, Clinical Significance, and Natural History of Left Ventricular Apical Aneurysms in Hypertrophic Cardiomyopathy

Martin S. Maron, MD; John J. Finley, MD; J. Martijn Bos, MD;



SD or ICD: 25%

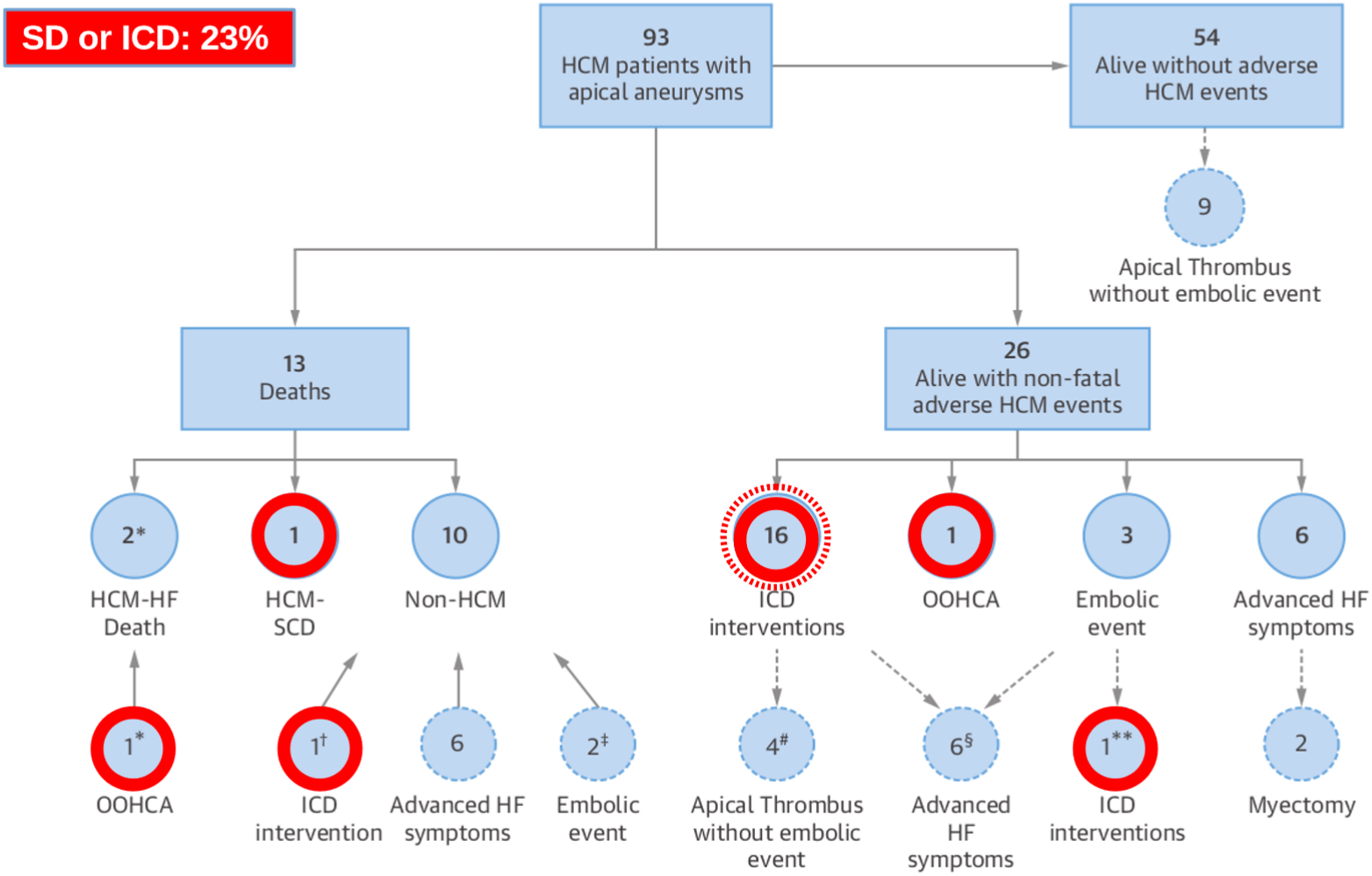
3 patients out of 7 without any risk factors for Sudden Death

Hypertrophic Cardiomyopathy With Left Ventricular Apical Aneurysm

Implications for Risk Stratification and Management

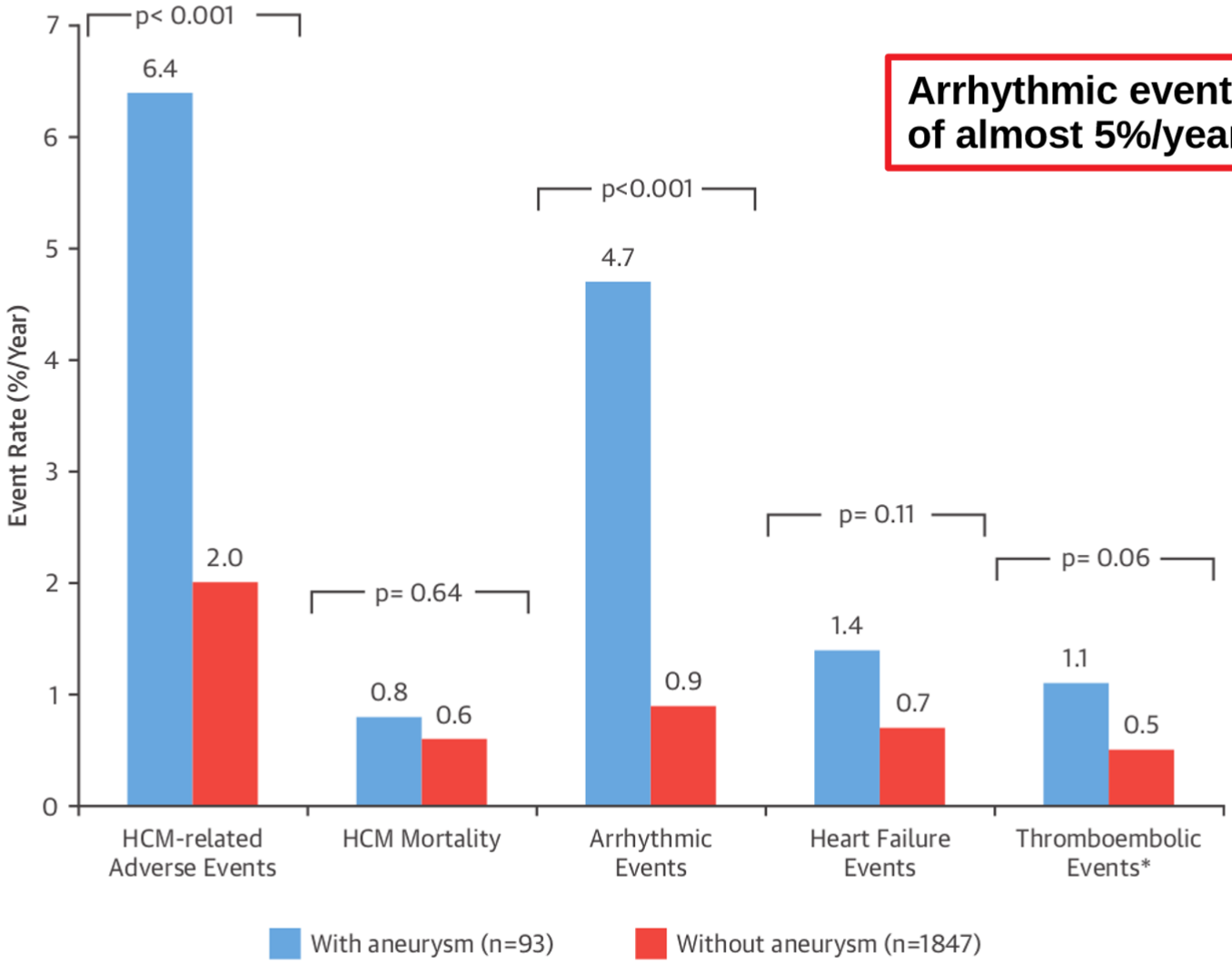
Ethan J. Rowin, MD,[a] Barry J. Maron, MD,[a] Tammy S. Haas, RN,[b] Ross F. Garberich, MS,[b] Weijia Wang, MD, Mark S. Link, MD,[a] Martin S. Maron, MD[a]

SD or ICD: 23%



Hypertrophic Cardiomyopathy

Event rate with or without apical aneurysm



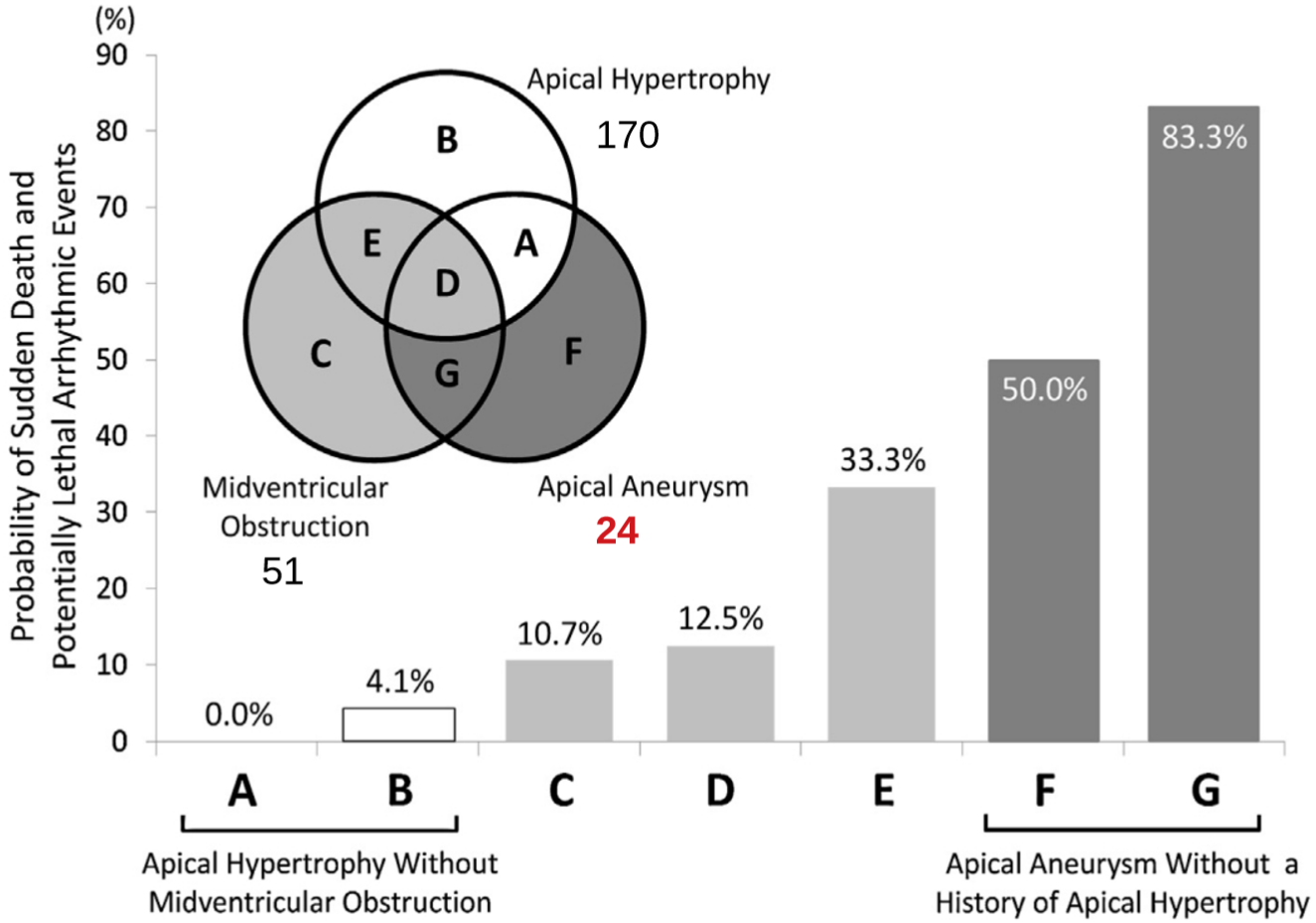
Arrhythmic event rate of almost 5%/year

Phenotypic overlap in hypertrophic cardiomyopathy: Apical hypertrophy, midventricular obstruction, and apical aneurysm

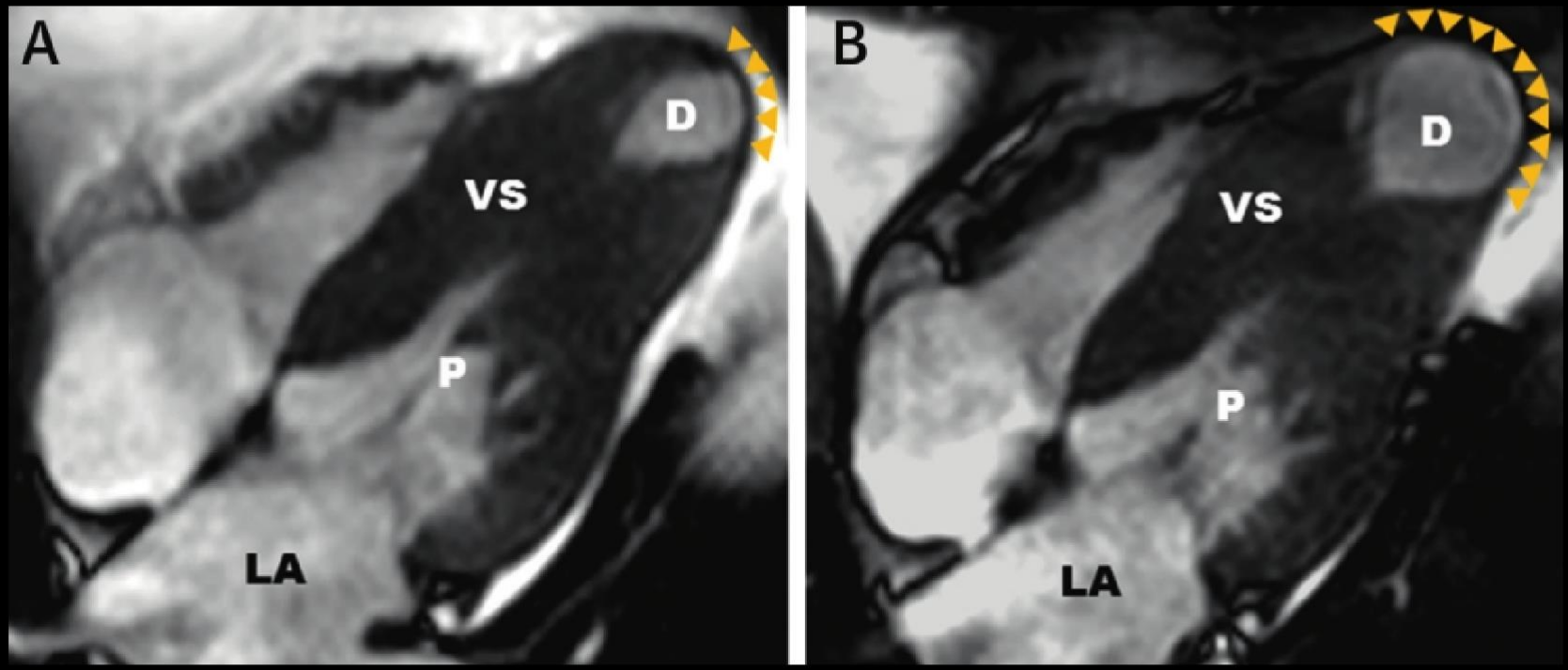
Yuichiro Minami (MD)*, Shintaro Haruki (MD), Nobuhisa Hagiwara (MD, FJCC)

Department of Cardiology, Tokyo Women's Medical University, Tokyo, Japan

Probability of Sudden Death / Lethal arrhythmias



Apical Aneurysm in Hypertrophic Cardiomyopathy



8 years later

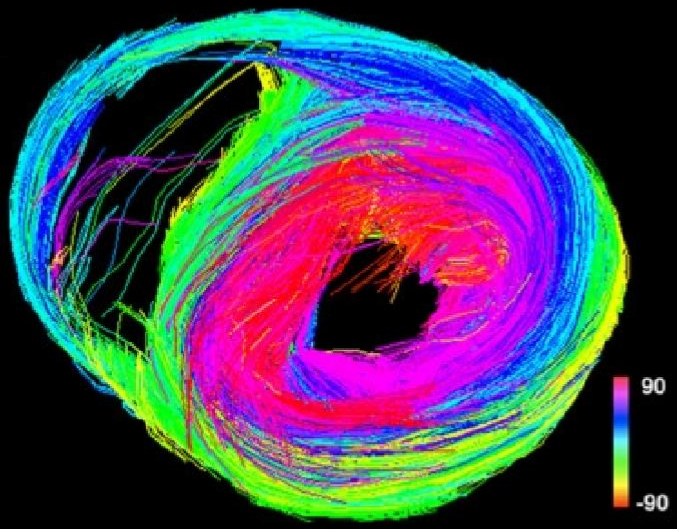
The ventricular myocardium, both right and left, exists as a continuous muscle band

Myofibers orientation

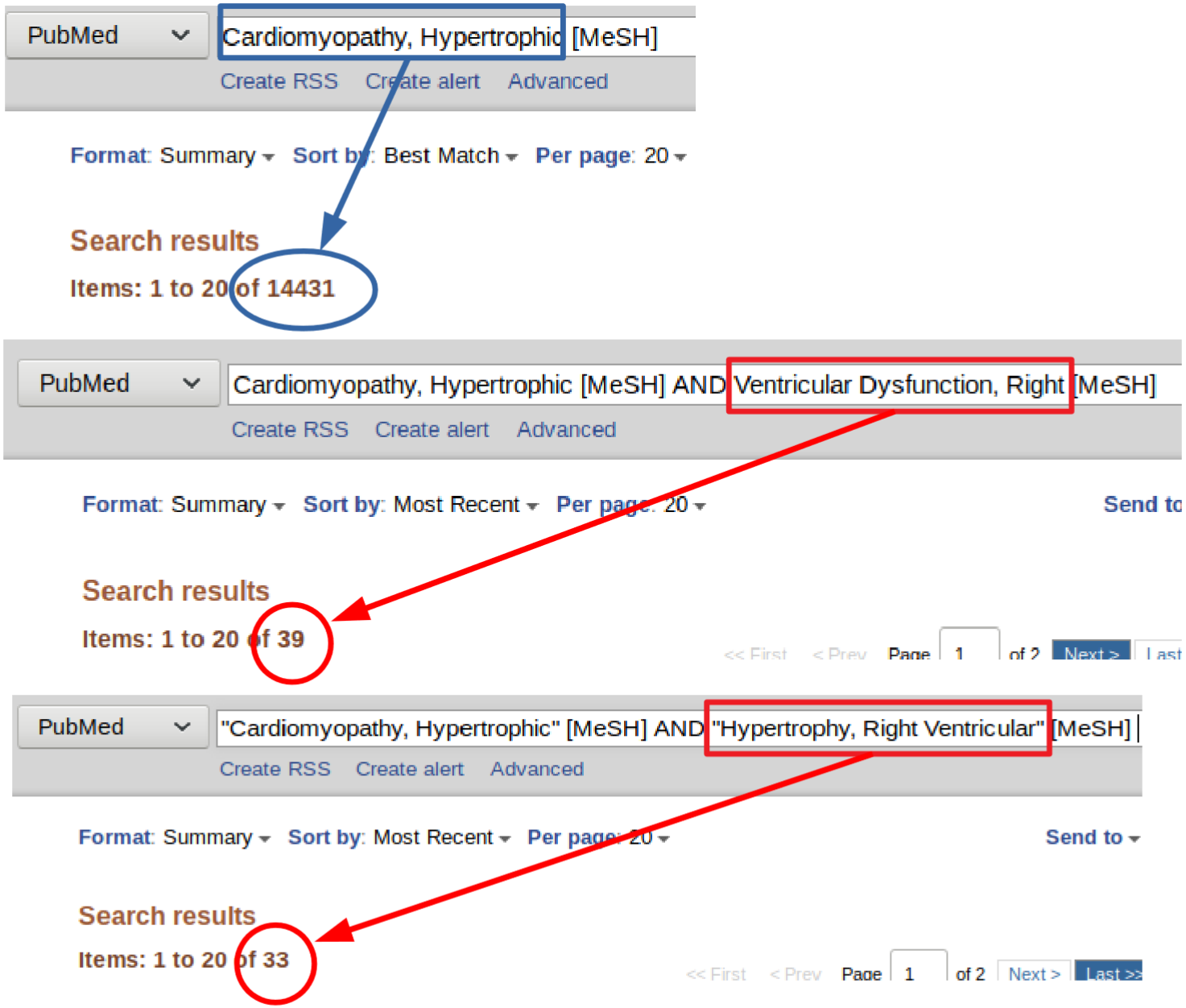
Red: clockwise

Green: counterclockwise

Blue: Circular



Diffusion Tensor Magnetic Resonance Imaging (DTMRI)


PubMed
Cardiomyopathy, Hypertrophic [MeSH]
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Create alert
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Format: Summary
Sort by: Best Match
Per page: 20
Search results
Items: 1 to 20 of 14431
PubMed
Cardiomyopathy, Hypertrophic [MeSH] AND Ventricular Dysfunction, Right [MeSH]
Create RSS
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Format: Summary
Sort by: Most Recent
Per page: 20
Send to
Search results
Items: 1 to 20 of 39
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PubMed
"Cardiomyopathy, Hypertrophic" [MeSH] AND "Hypertrophy, Right Ventricular" [MeSH]
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Send to
Search results
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Page 1 of 2
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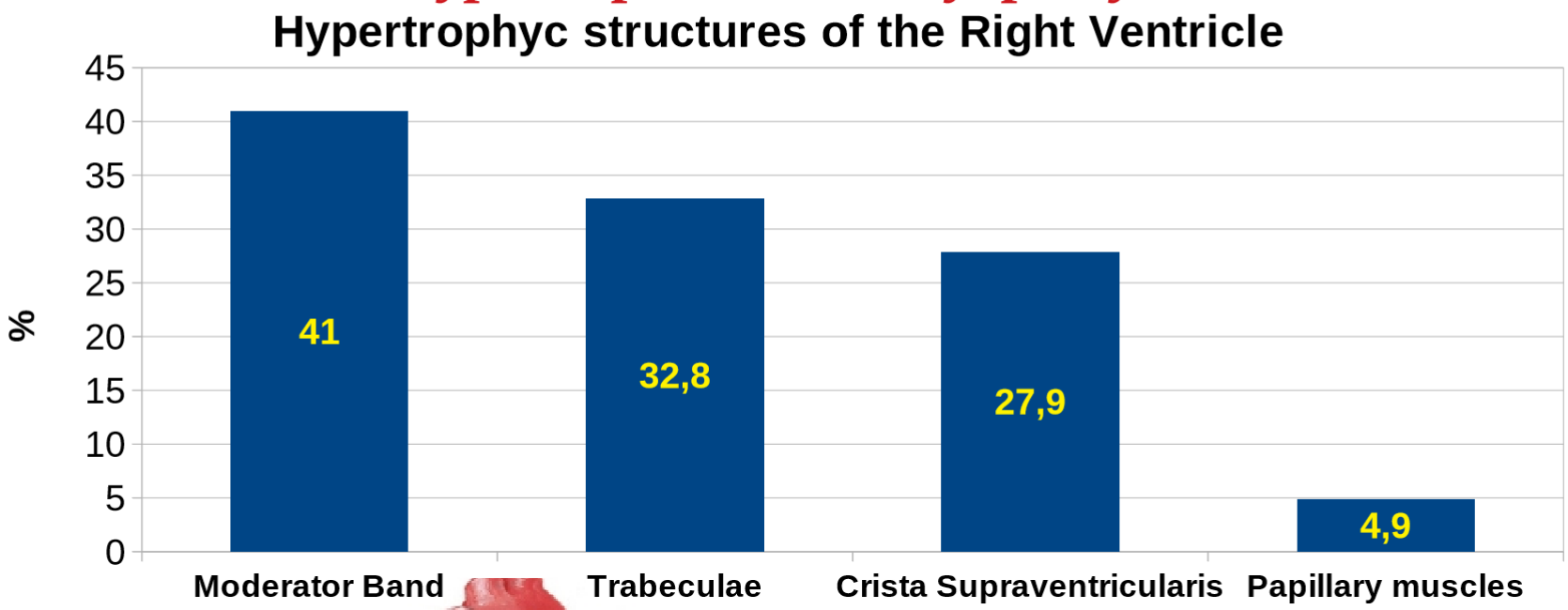
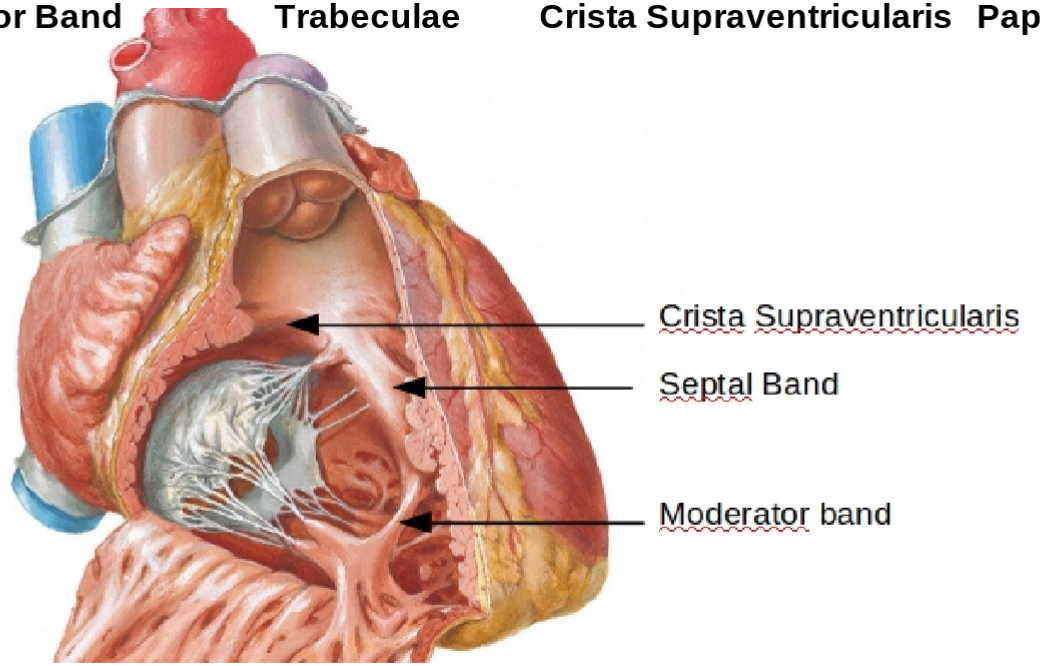
Right Ventricular Hypertrophy in Hypertrophic Cardiomyopathy

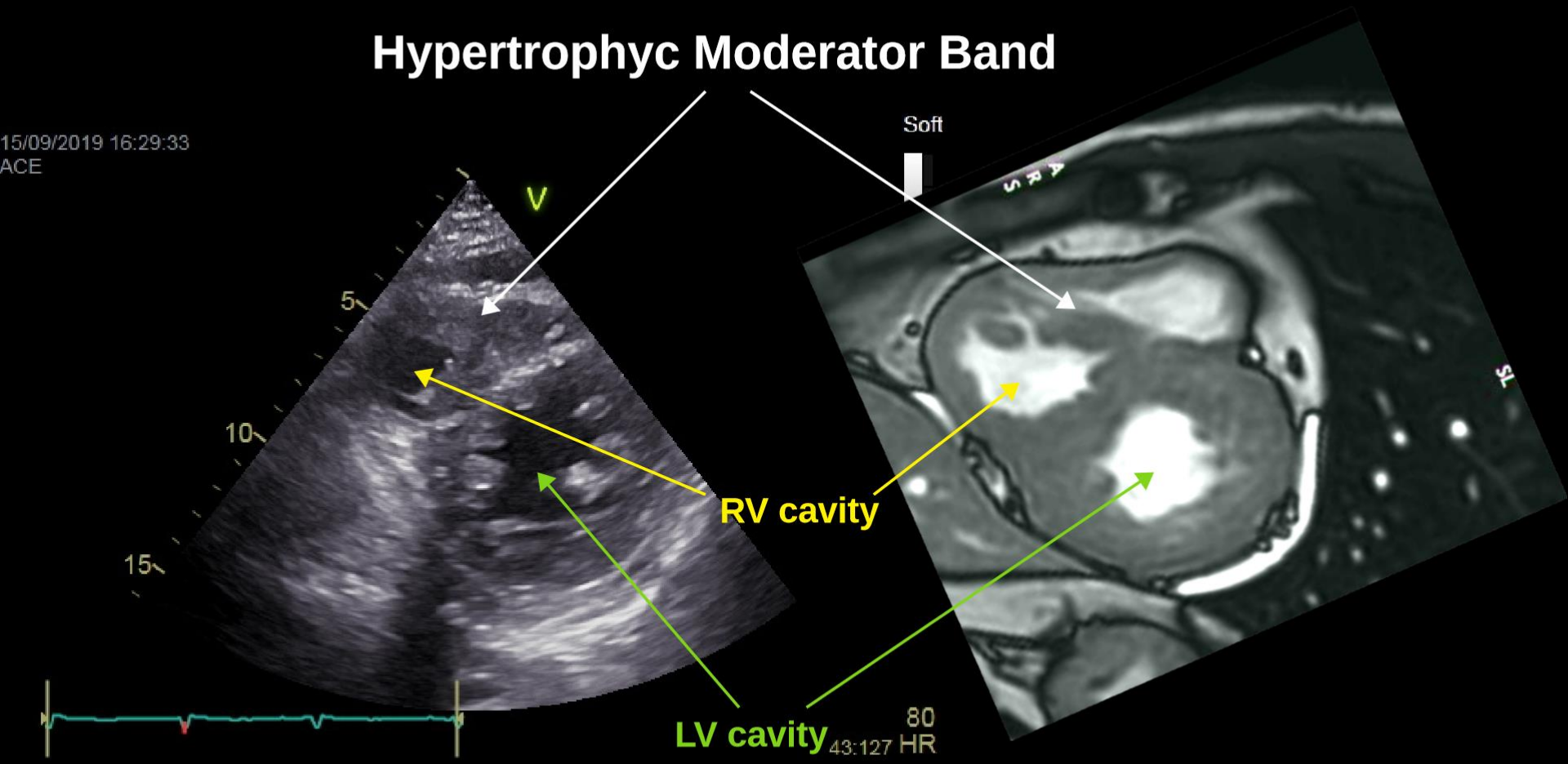
Autor	*year*	*N° of patients*	*Patients with RVH*	*notes*
McKenna	1988	73	**44%**	Echo ≥6 mm
Roșca	2015	99	**53%**	Echo >5mm
Maron	2007	46	**33%**	CMR 2SD>5±1 mm
Nagata	2015	106	**28%**	CMR >5 mm
Our cases Merello	2019	61	**38% RV free wall 62% other RV structures**	CMR ≥5 mm

Hypertrophic Cardiomyopathy



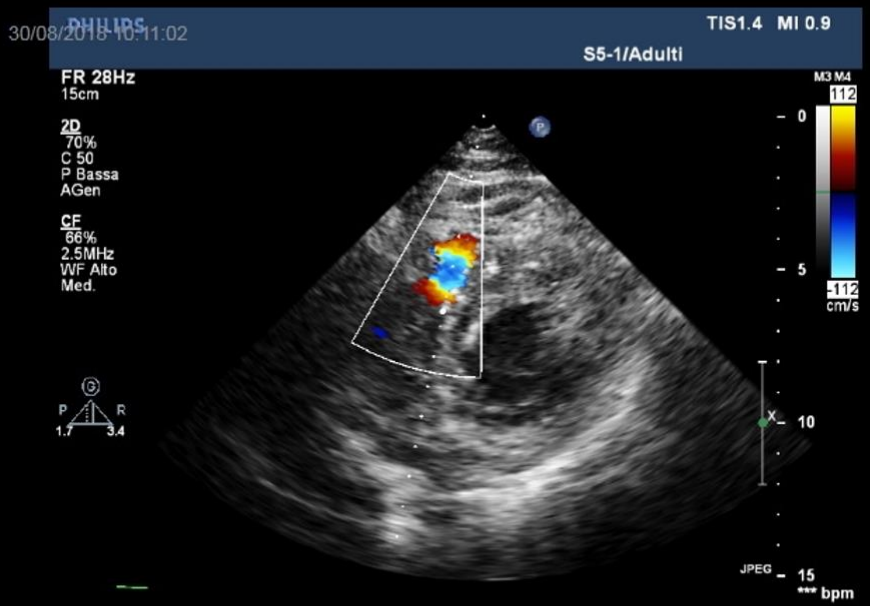


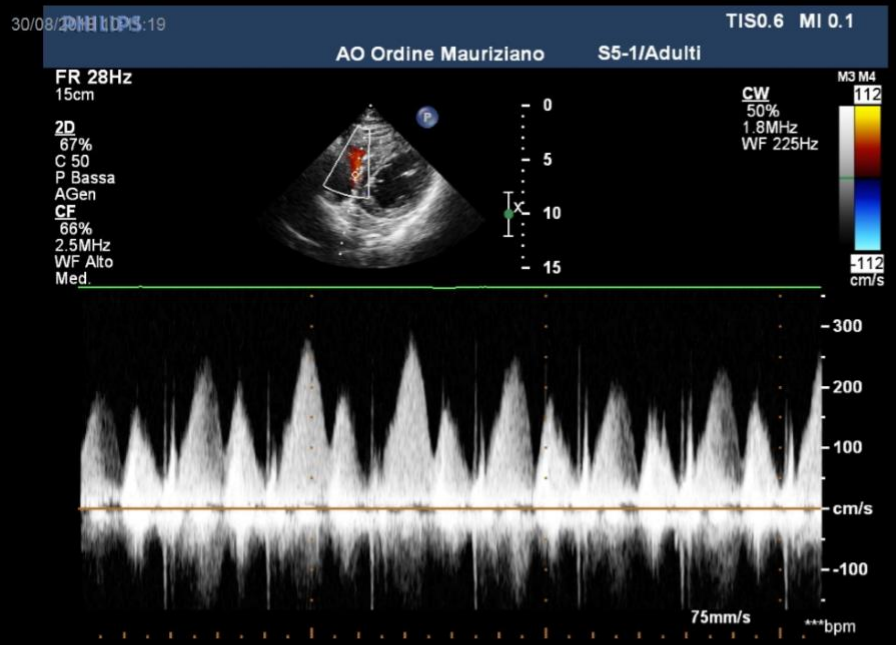
Hypertrophic Cardiomyopathy



Hypertrophic Cardiomyopathy

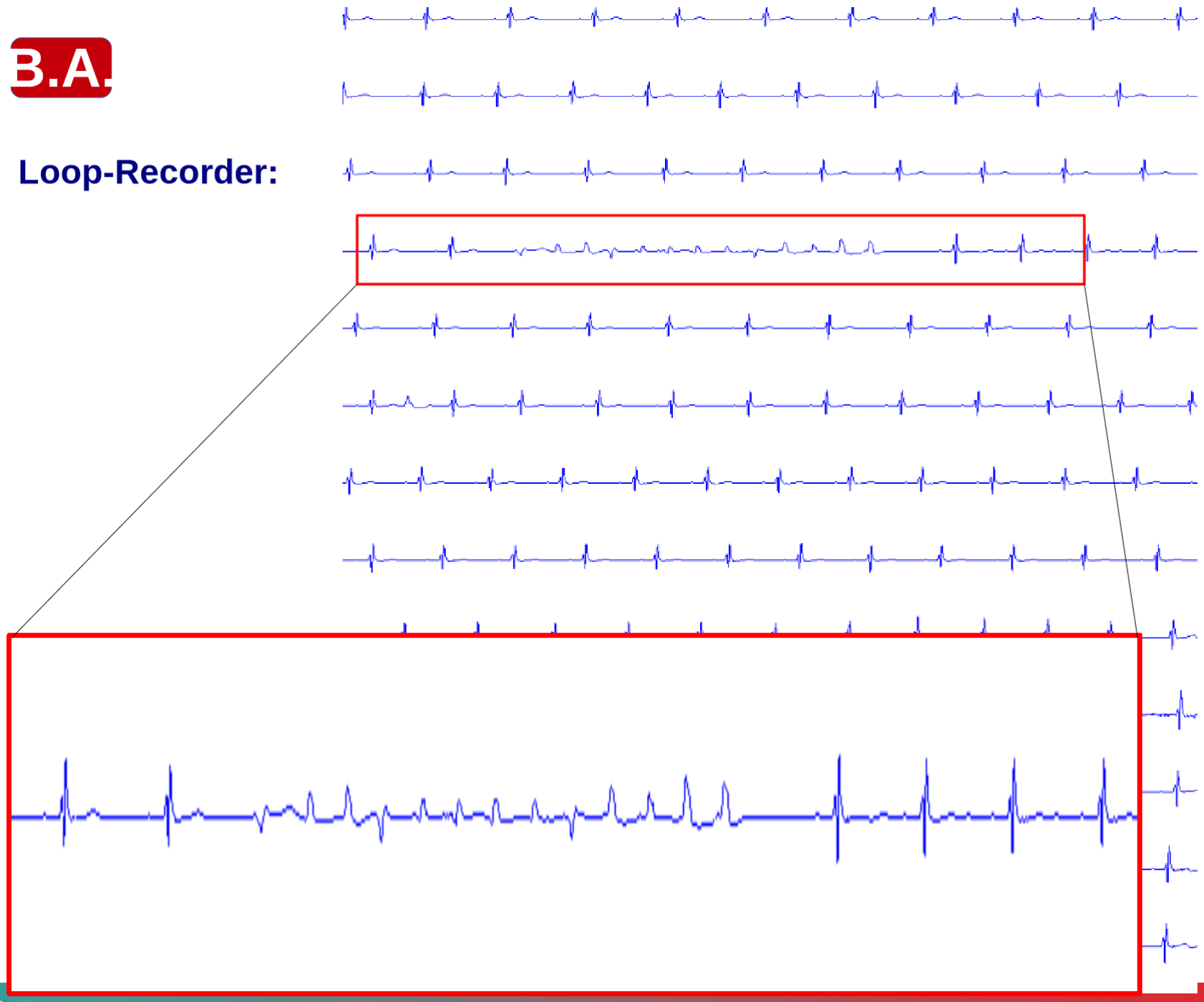
Flow turbulence and gradient at level of the Hypertrophic Moderator Band





B.A.

Loop-Recorder:

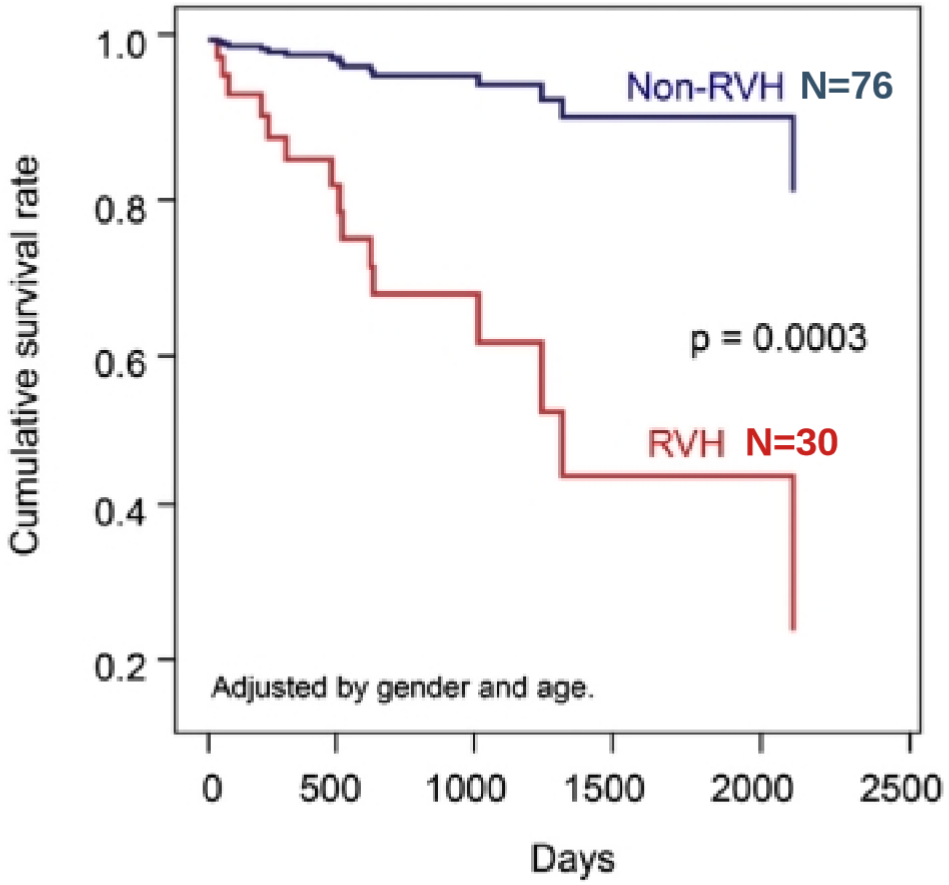


Right Ventricular Remodeling, Its Correlates, and Its Clinical Impact in Hypertrophic Cardiomyopathy

Table 5 Correlates of ventricular arrhythmia in patients with HCM (N=99)

Variables	Univariate analysis			Multivariate analysis
	OR	95% CI	*P*	*P*
Age	0.973	0.942–1.006	.10	.25
LAVi	0.996	0.974–1.019	.75	—
Septal S	0.891	0.632–1.255	.50	—
Lateral S	0.782	0.536–1.142	.20	—
Mean e′	0.899	0.662–1.222	.49	—
E/e′ ratio	0.970	0.881–1.069	.53	—
LVMi	1.003	0.995–1.012	.45	—
LVWT	1.166	1.039–1.308	.009	.17
LV GLS	1.182	0.996–1.402	.05	.35
RVWT (N=52)	1.850	1.314–2.605	<.001	.002
RV S velocity	0.902	0.730–1.113	.33	—
RV GLS	1.069	0.936–1.221	.32	—
RVWε	1.014	0.915–1.124	.78	—
sPAP	0.980	0.925–1.038	.48	—
LV outflow tract gradient	1.000	0.971–1.030	.98	—
MR degree	0.900	0.493–1.643	.73	—

Right Ventricular Hypertrophy Is Associated With Cardiovascular Events in Hypertrophic Cardiomyopathy: Evidence From Study With Magnetic Resonance Imaging



(hazard ratio, 5.42; 95% IC, 1.16-25.3)

Hypertrophic Cardiomyopathy

Take  message

- Measure Left Atrium
- Look for apical aneurysm and follow it up
- Check the right ventricle

Thank you