Risk stratification for SCD in HCM pts

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Review

How Hypertrophic Cardiomyopathy Became a Contemporary Treatable Genetic Disease With Low Mortality
Shaped by 50 Years of Clinical Research and Practice

Barry J. Maron, MD; Ethan J. Rowin, MD; Susan A. Casey, RN; Martin S. Maron, MD

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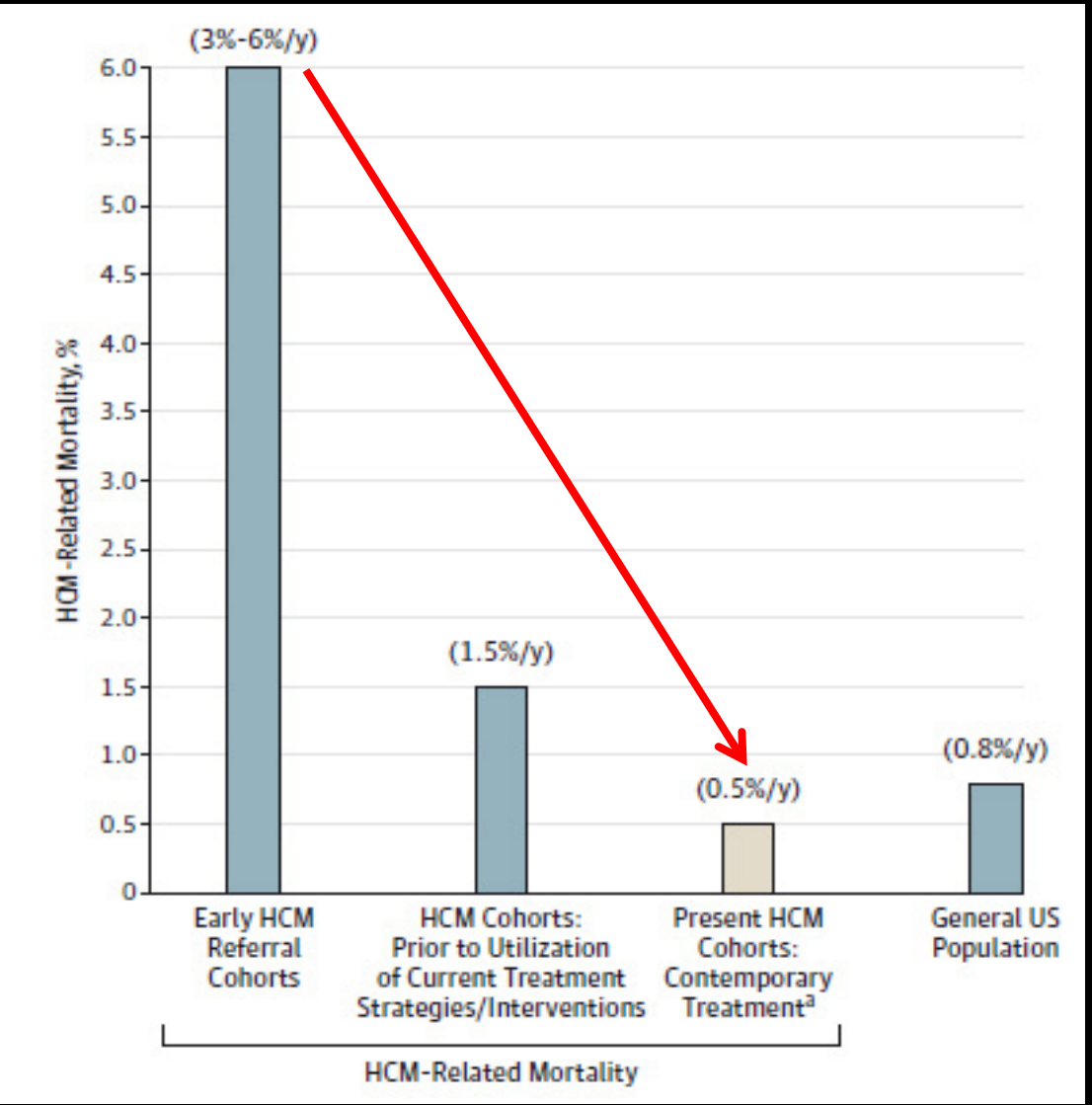

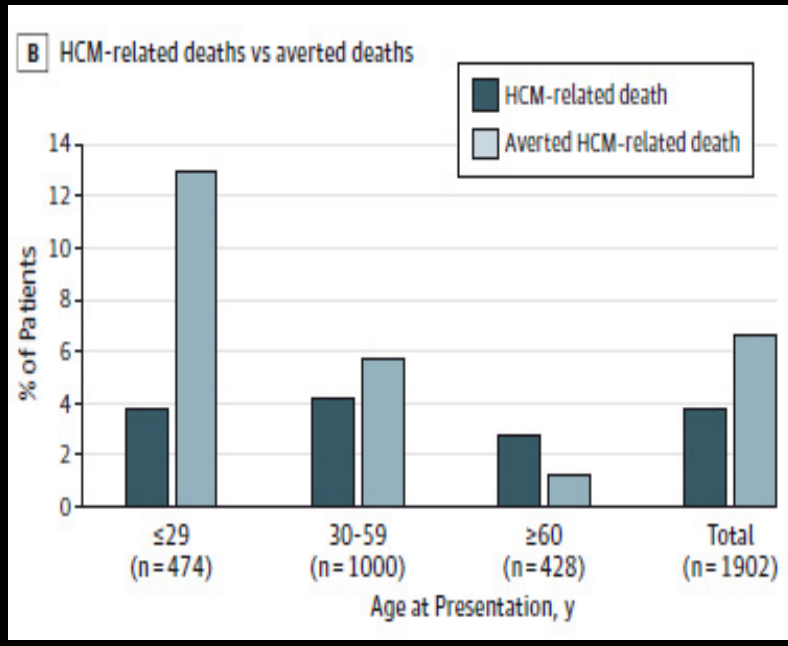

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How Hypertrophic Cardiomyopathy Became a Contemporary Treatable Genetic Disease With Low Mortality

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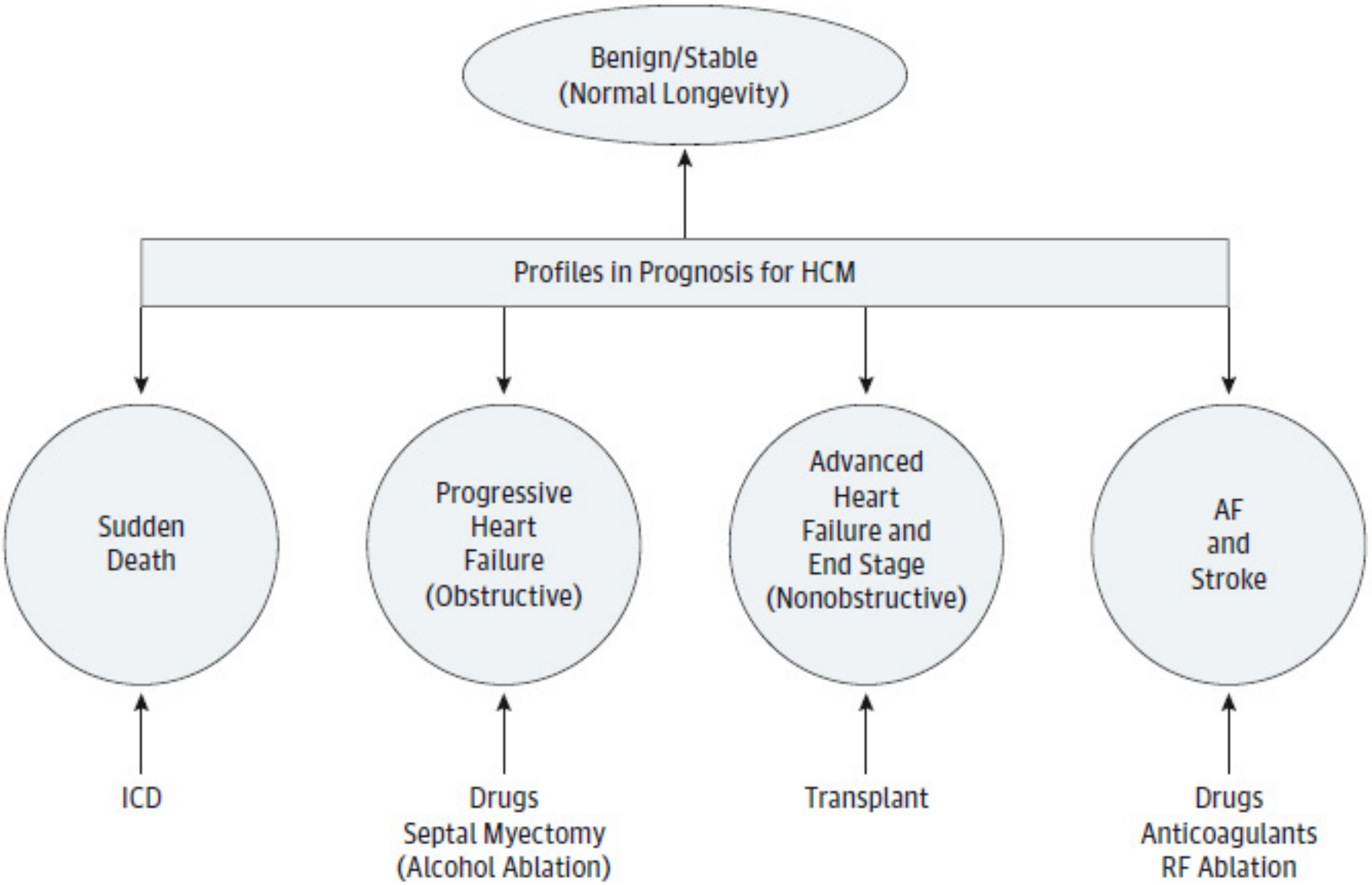
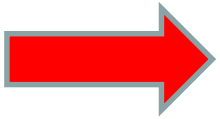


Invasive Electrophysiological Testing

Recommendations on electrophysiologic testing	Class[a]	Level[b]
Invasive electrophysiological study is recommended in patients with documented persistent or recurrent supraventricular tachycardia (atrial flutter, atrial tachycardia, atrioventricular nodal re-entry tachycardia, accessory atrioventricular pathway mediated tachycardias) and in patients with ventricular pre-excitation, in order to identify and treat an ablatable substrate.	I	C
Invasive electrophysiological study may be considered in selected patients with documented, symptomatic, monomorphic, sustained (>30 s) ventricular tachycardia in order to identify and treat an ablatable arrhythmia substrate.	IIb	C
Invasive electrophysiological study with programmed ventricular stimulation is not recommended for sudden cardiac death risk stratification.	III	C

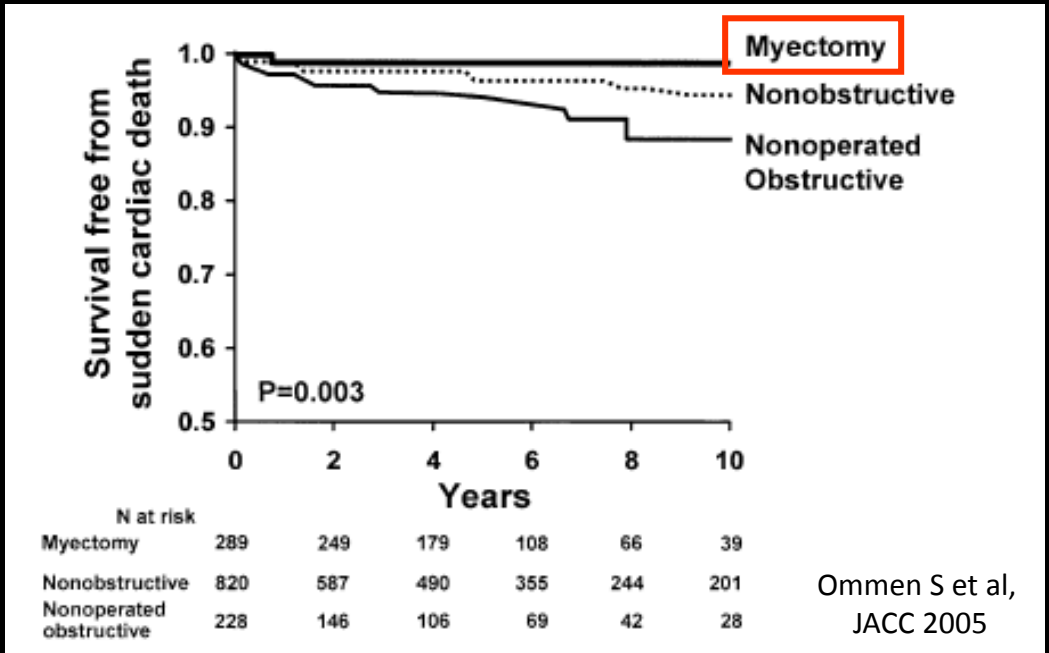


Lifestyle and treatment options for SCD risk reduction

- Avoidance of competitive sports is recommended (Class I)
- AF ablation for AF with rapid ventricular response
- Myectomy reduces SD risk in HOCM pts

SURVIVAL FREE OF SUDDEN DEATH

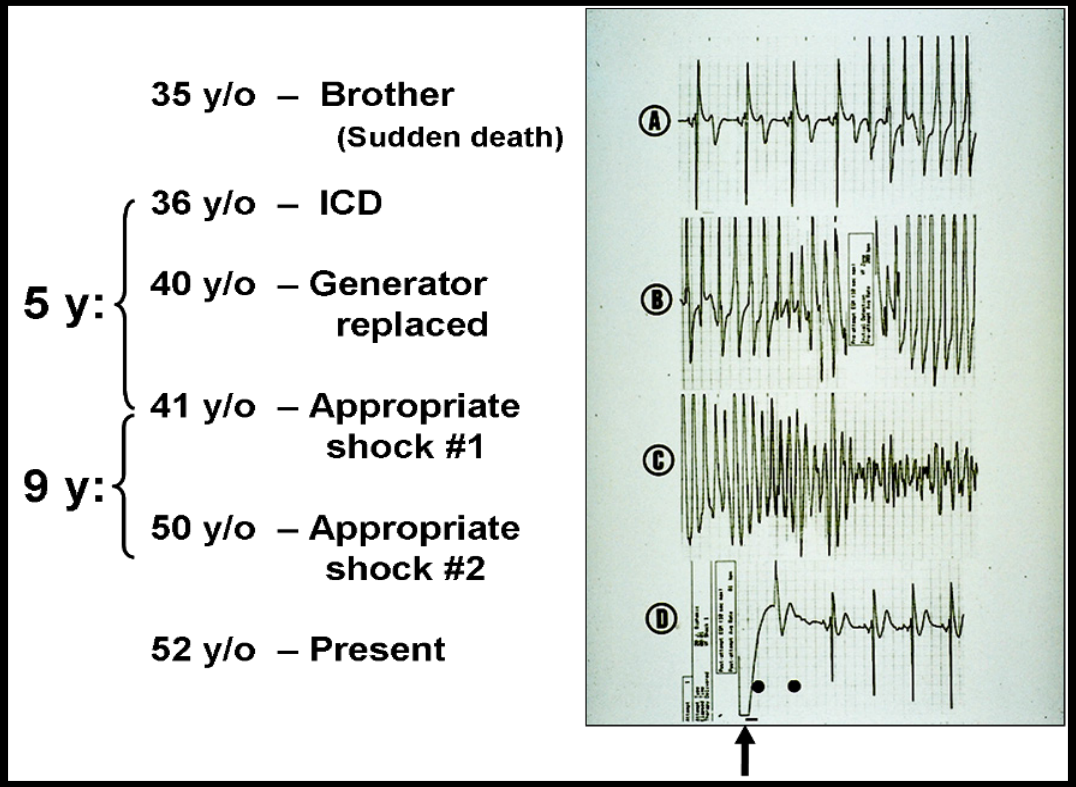
(Rochester Mayo Clinic, versus Minneapolis + Florence)



Ommen S et al, JACC 2005

HCM and UNEXPECTED SUDDEN CARDIAC DEATH : FACTS

- UNEXPECTED SCD is a catastrophe
- It is a random event , fortunately rare, which may occur at any time
- Its incidence decreases later in life
- VF is usually, but not always, the causative arrhythmia
- VF may be preceded by sinus tachicardia, AF or sustained VT



RISK STRATIFICATION: RISK FACTORS

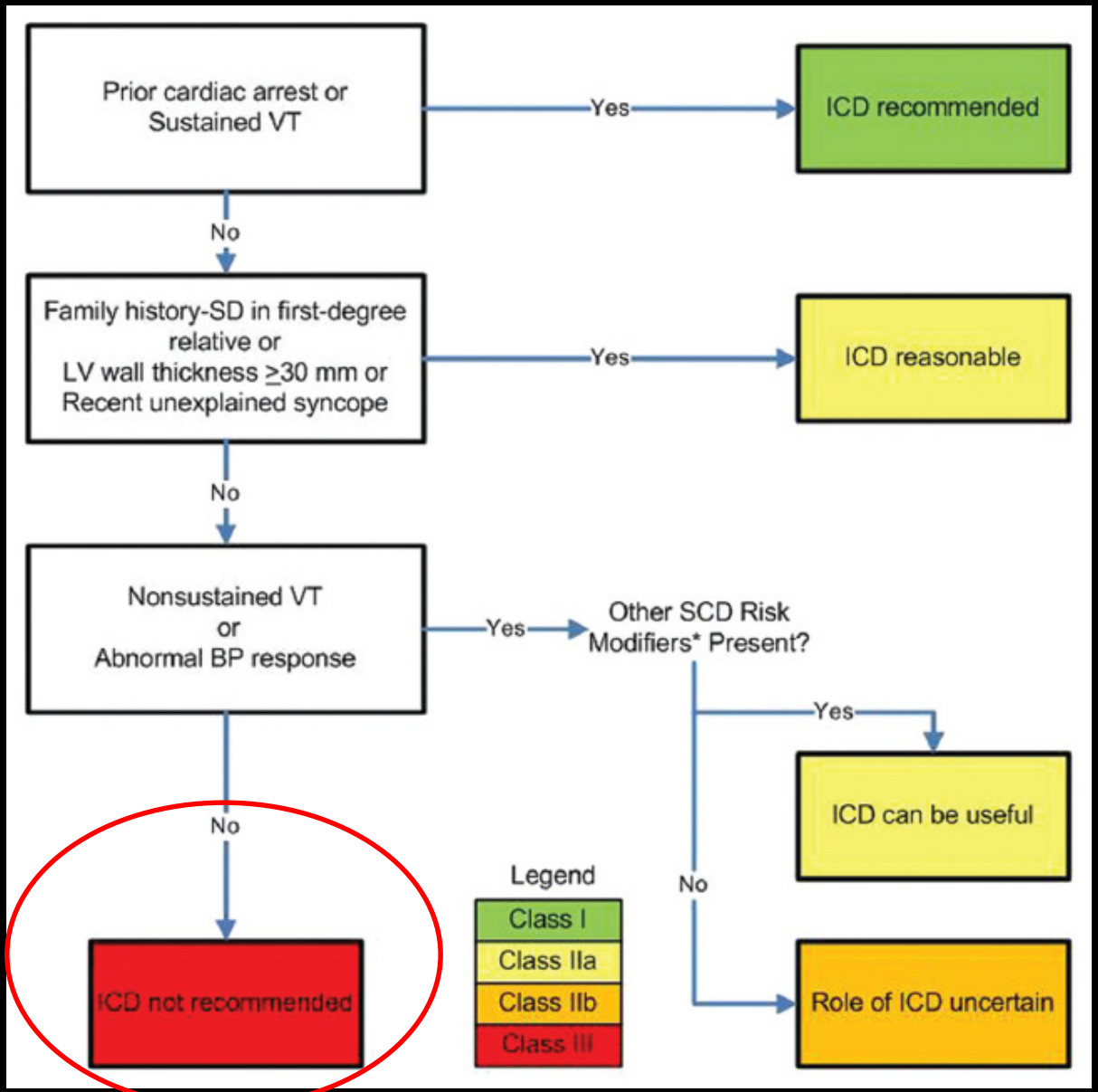
(ICD Guidelines for HCM AHA/ESC 2003 & AHA 2011)

RISK FACTOR	Sensitivity	Specificity	PPV	NPV
Family history of SD	42	79	28	88
Max LV thickness >30 mm	26	88	13	95
NSVT run > 120’	69	80	22	97
Abnormal pressure response at exercise test age < 45	75	66	15	97
Syncope	29	83	25	86

Elliot P, W.McKenna, 2003

2011 ACCF/AHA Guideline for the Diagnosis and Treatment of Hypertrophic Cardiomyopathy

A Report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines



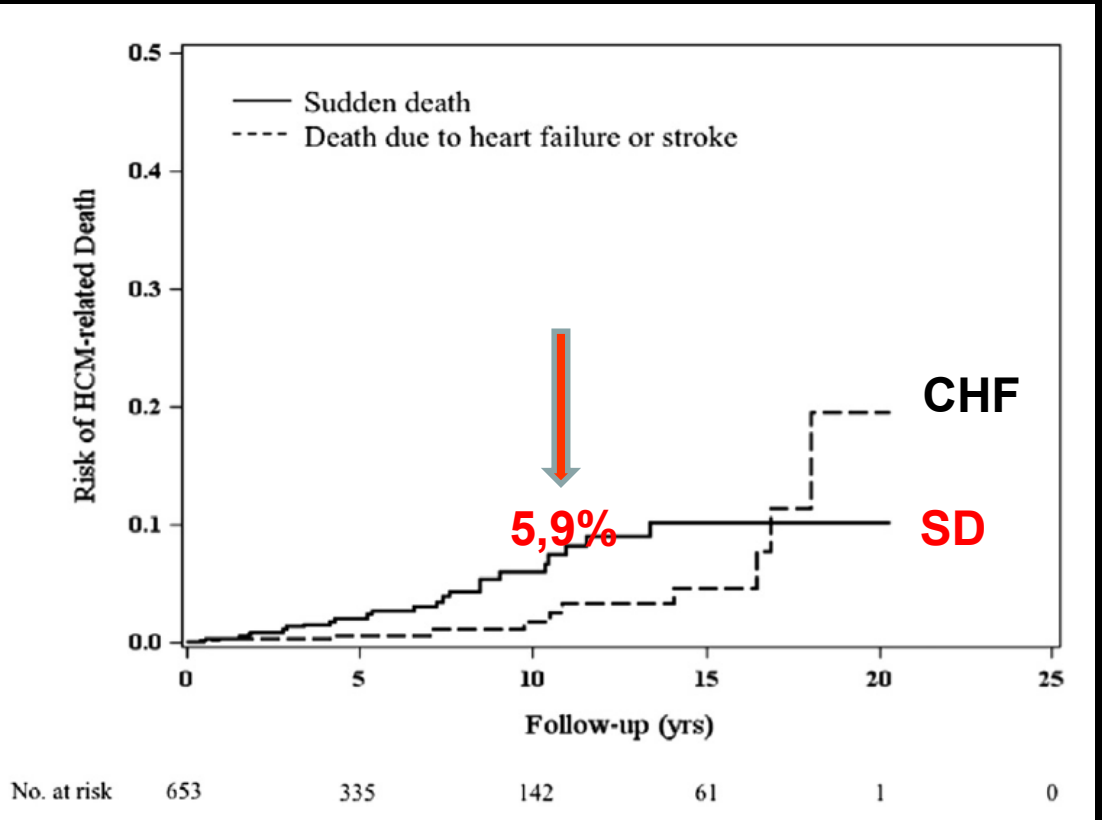
Risk of Sudden Death and Outcome in Patients With Hypertrophic Cardiomyopathy With Benign Presentation and Without Risk Factors

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[g], Beatrice Musumeci, MD[h], Francesco Lai, MD[e], Laura Yeates, BSc[d], Maria Rosa Conte, MD[g], Claudio Rapezzi, MD[c], Luca Boni, MD[g], and Barry J. Maron, MD[d]

Am J Cardiol 2014;113:1550–1555

THE «NO» RISK FACTOR COHORT

These findings demonstrate that identification of the individual patient with HC at increased risk of SD remains incomplete across the broad HC spectrum, including patients currently judged to be at low risk, and underscore the need for a more precise characterization of the risk profile and outcome of patients with HC.



RF for SD at multivariate analysis

Age	**0.97**	(0.94-0.99)
LA 41-50 mm	**3.11**	(1.13-8.54)
LA > 50 mm	**8.01**	(2.08-30.9)

Individual SCD risk assessment in HCM pts in clinical practice

1. Can I predict the risk of SCD in 5 years ?
2. Can I reduce the risk without implanting an ICD ?
3. If I feel he/she needs an ICD, which one ?
4. If I feel he/she needs an ICD, will the patient agree ?

A novel clinical risk prediction model for sudden cardiac death in hypertrophic cardiomyopathy (HCM Risk-SCD)

Constantinos O'Mahony[1], Fatima Jichi[2], Menelaos Pavlou[8], Lorenzo Monserrat[3], Aristides Anastasakis[4], Claudio Rapezzi[5], Elena Biagini[5], Juan Ramon Gimeno[6], Giuseppe Limongelli[7], William J. McKenna[1], Rumana Z. Omar[2,8] and Perry M. Elliott[1]*, for the Hypertrophic Cardiomyopathy Outcomes Investigators

European Heart Journal 2013

Retrospective multicenter study

(6 centers)

3675 pts age > 16 yrs at diagnosis

Predictor variable	SCD risk prediction model	
	Hazard ratio (95% confidence interval)	*P*-value
Age (years)	0.98 (0.97, 0.99)	0.001
Maximal wall thickness (mm)	1.17 (1.01, 1.37)	0.042
Left atrial diameter (mm)	1.03 (1.01, 1.05)	0.006
LV outflow gradient (mmHg)	1.004 (1.001, 1.01)	0.021
Family history SCD	1.58 (1.18, 2.13)	0.002
NSVT	2.29 (1.64, 3.18)	<0.001
Unexplained syncope	2.05 (1.48, 2.82)	<0.001

Incomplete dataset for Abnormal blood pressure response at exercise

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)

http://www.escardio.org/Guidelines-&-Education/

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

Risk of SCD at 5 years (%):

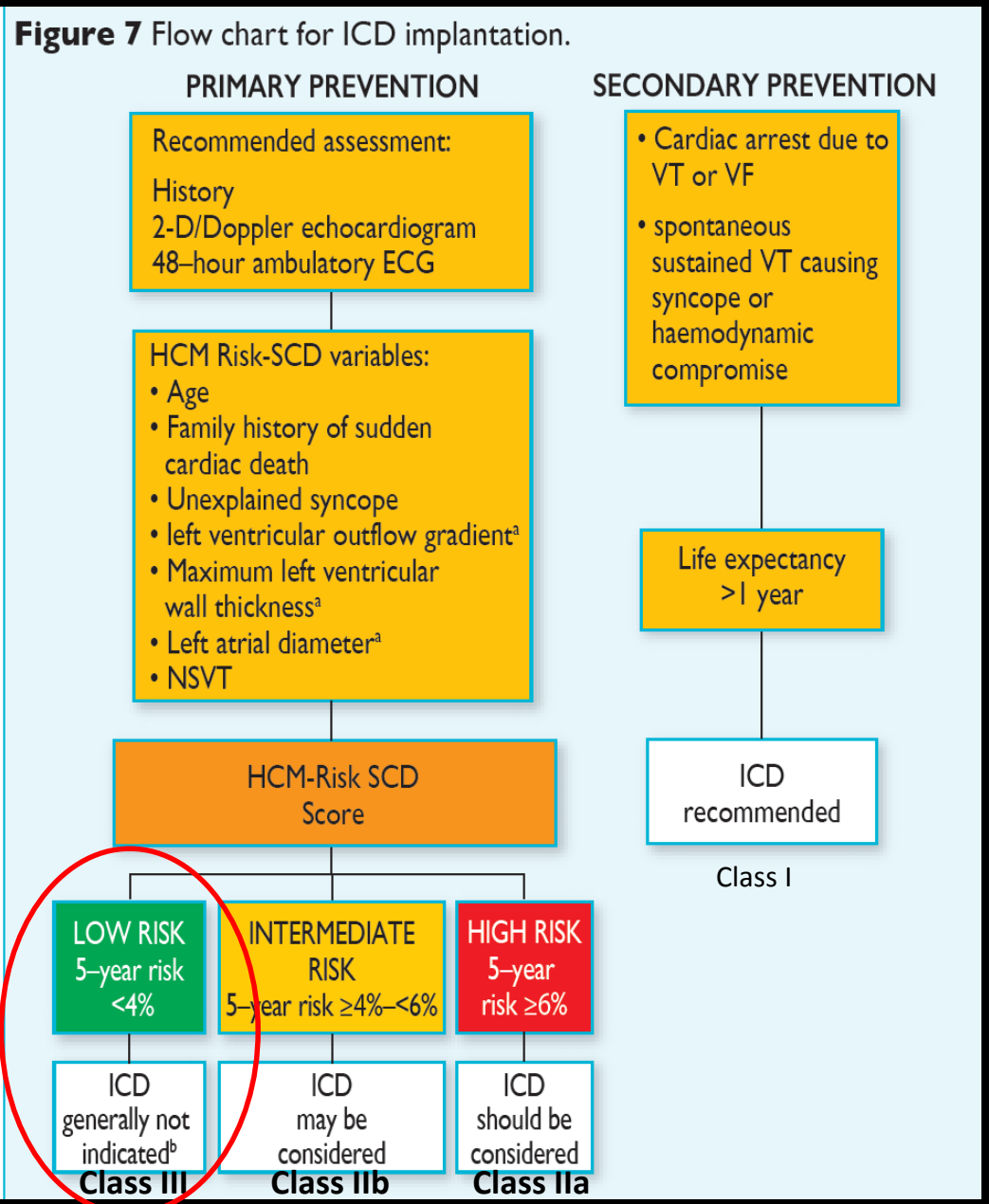
HCM Risk-SCD Calculator

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.*

It is recommended that the 5-year risk of SCD be assessed at first evaluation and re-evaluated at 1–2 year intervals or whenever there is a change in clinical status (Class I)

2014 ESC Guidelines on diagnosis and management of hypertrophic cardiomyopathy
European Heart Journal
doi:10.1093/eurheartj/ehu284

Figure 7 Flow chart for ICD implantation.



Recommendations for ICD in each risk category take into account not only the absolute statistical risk,

but also age and

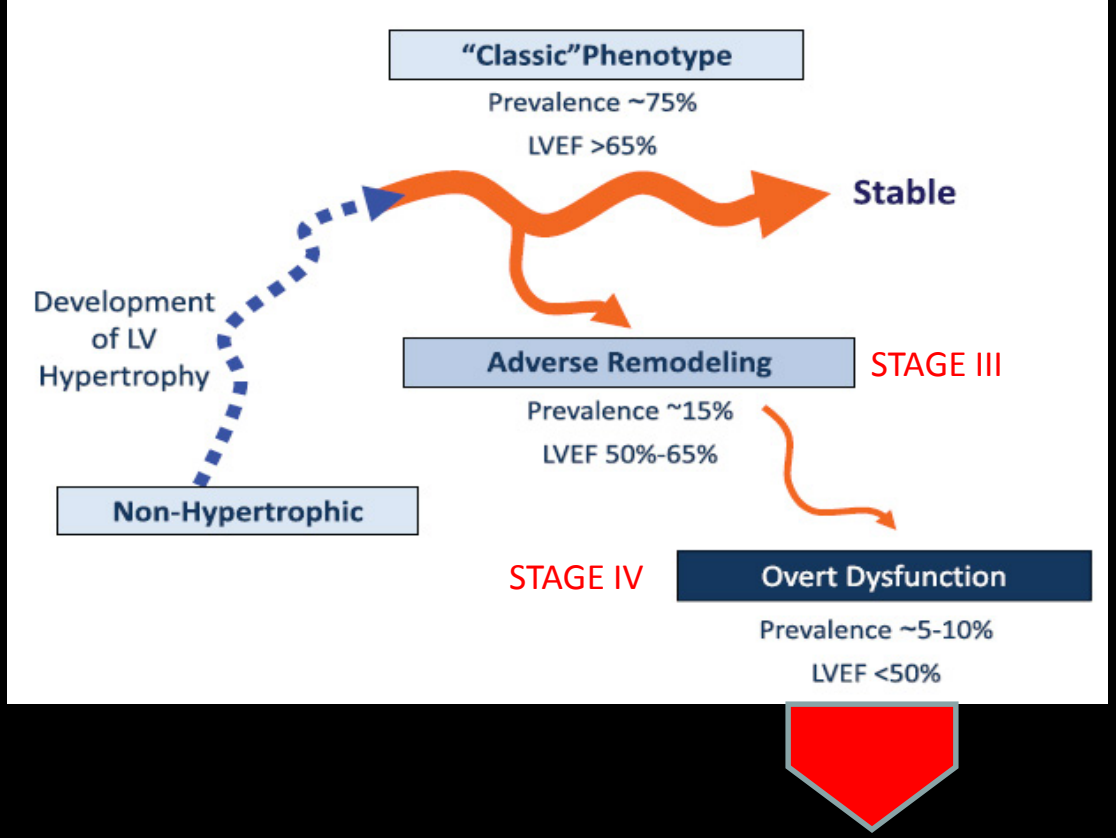
- general health of the patient,
- socio-economic factors and the
- psychological impact of therapy

Patterns of Disease Progression in Hypertrophic Cardiomyopathy

An Individualized Approach to Clinical Staging

Iacopo Olivotto, MD; Franco Cecchi, MD; Corrado Poggesi, MD; Magdi H. Yacoub, MD, FRS

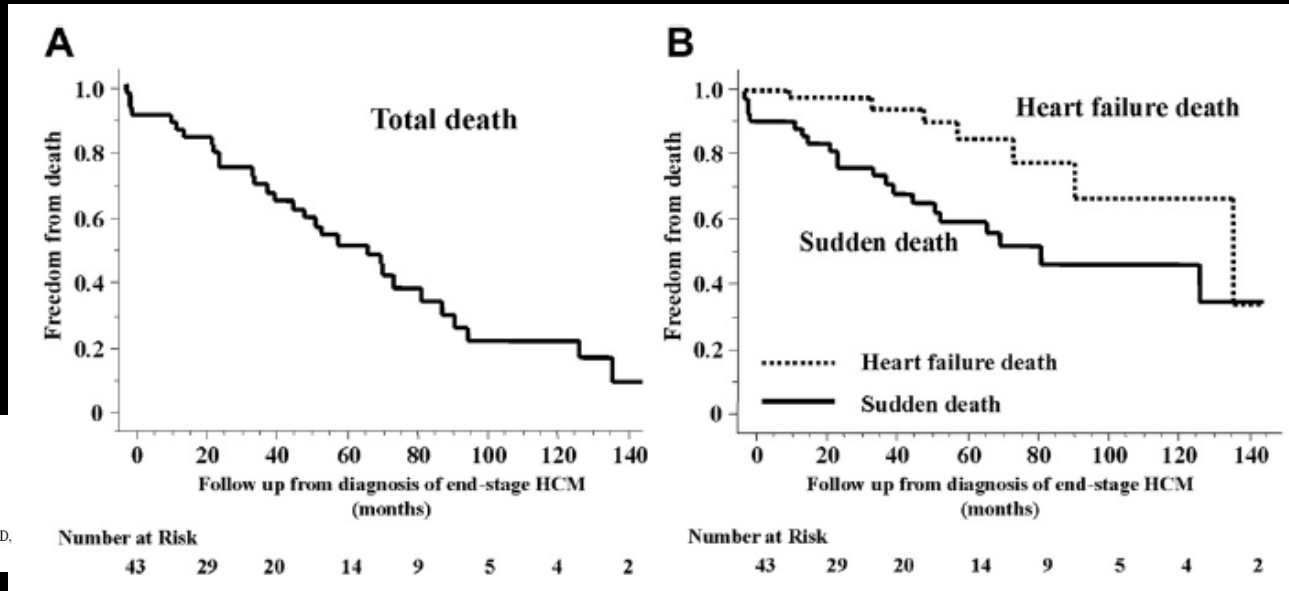
Circ Heart Fail. 2012;5:535-546



Risk of Sudden Death in End-Stage Hypertrophic Cardiomyopathy

HIROTAKA KAWARAI, MD, KATSUYA KAJIMOTO, MD, YUICHIRO MINAMI, MD, NOBUHISA HAGIWARA, MD, AND HIROSHI KASANUKI, MD

J Cardiac Fail 2011;17:459–464



Additional Risk Factors for SCD risk stratification

- CHF : end stage /overt dysfunction disease (Stage III-IV)
- CMR : Extent of LGE (>15%)
- EX TEST : Abn BP response / Ventricular arrhythmias (NSVT /VF)
- ECG : pseudo STEMI pattern
- Cardiopulmonary ex test : VE/VCO2 > 31
- ECHO/CMR : Midventricular obstruction with apical aneurism

Usefulness of Electrocardiographic Patterns at Presentation to Predict Long-term Risk of Cardiac Death in Patients With Hypertrophic Cardiomyopathy

Elena Biagini, MD, PhD[a], Chiara Pazzi, MD[a], Iacopo Olivotto, MD[b], Beatrice Musumeci, MD[c], Giuseppe Limongelli, MD[d], Giuseppe Boriani, MD, PhD[a], Giuseppe Pacileo, MD[d], Vittoria Mastromarino, MD[c], Maria Letizia Bacchi Reggiani, BSc[a], Massimiliano Lorenzini, MD[a], Francesco Lai, MD[a], Alessandra Berardini, MD[a], Francesca Mingardi, MD[a], Stefania Rosmini, MD, PhD[a], Elvira Resciniti, MD[a], Claudia Borghi, MD, PhD[a], Camillo Autore, MD[c], Franco Cecchi, MD[b], and Claudio Rapezzi, MD[a,*]

Am J Cardiol 2016

Studio multicentrico Bologna, Firenze, Roma, Napoli 1004 pz

Multivariate analysis in the overall population

Variables	SCD or surrogates Multivariable analysis		Major cardiovascular events Multivariable analysis	
	H.R. (95% CI)	P value	H.R. (95% CI)	P value
Age			1.02 (1.01 - 1.03)	0.0001
Unexplained syncope	2.47 (1.37 - 4.47)	0.003		
NSVT 24 hour Holter monitoring	1.68 (1.06 - 2.65)	0.027		
LV ejection fraction <50%	3.55 (1.89 - 6.66)	0.0001	3.73 (2.39 - 5.83)	0.0001
QRS duration ≥120 msec	1.78 (1.05 - 3.03)	0.033	1.69 (1.16 - 2.47)	0.007
Low QRS voltages	2.26 (1.01 - 5.07)	0.048		
"Pseudo STEMI" pattern	2.28 (1.38 - 3.77)	0.001	1.66 (1.13 - 2.45)	0.010
Prolonged QTc interval (≥440 msec)			1.68 (1.21 - 2.34)	0.002

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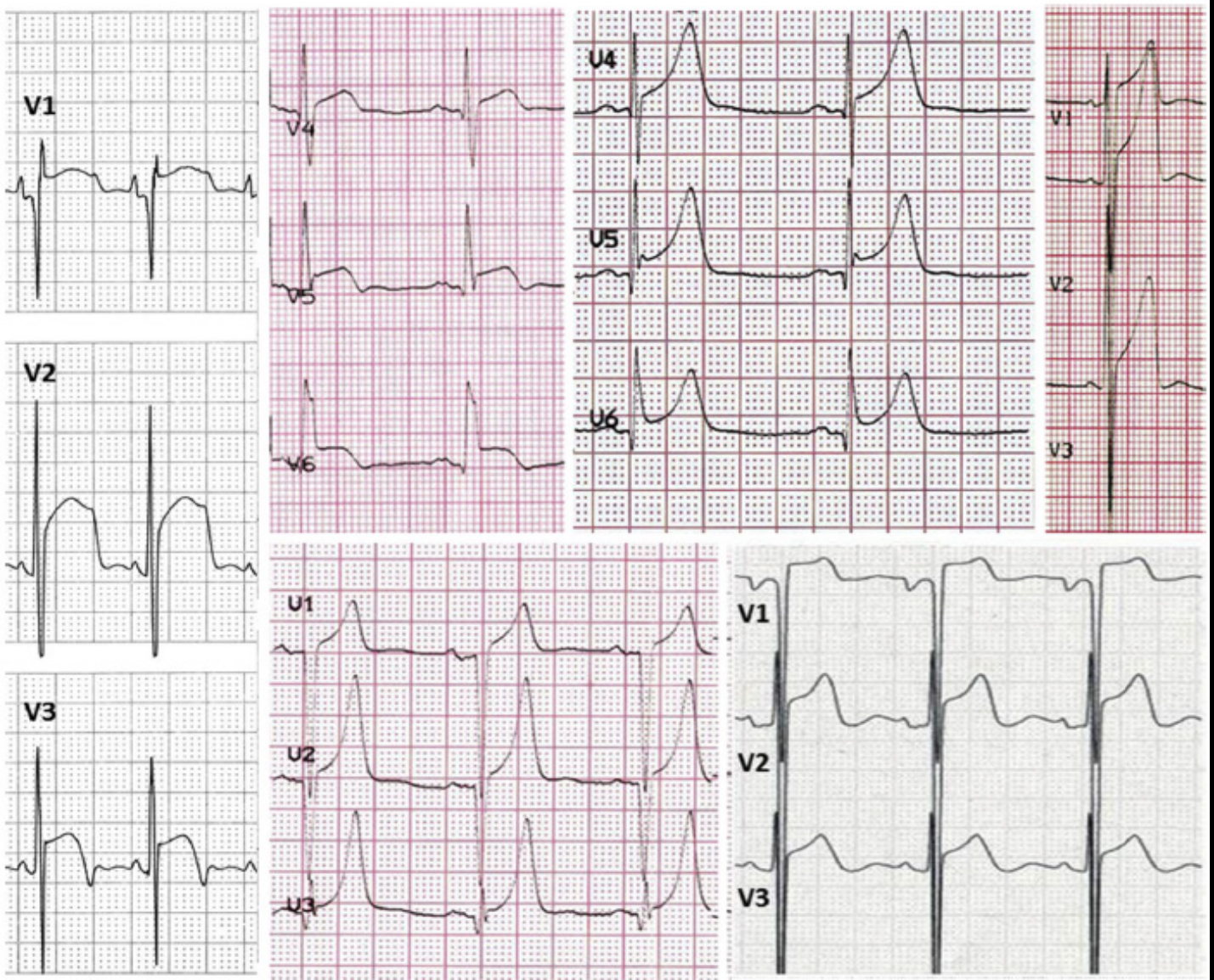
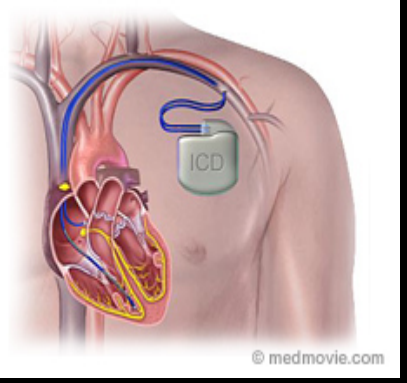
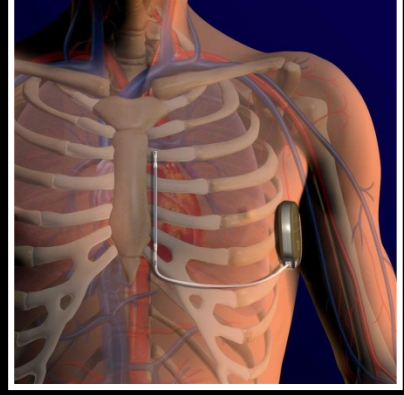
Cardiomyopathy/ECG Patterns and Cardiac Death in HC





CHOICE BETWEEN STANDARD VERSUS SUBCUTANEOUS ICD (S-ICD)

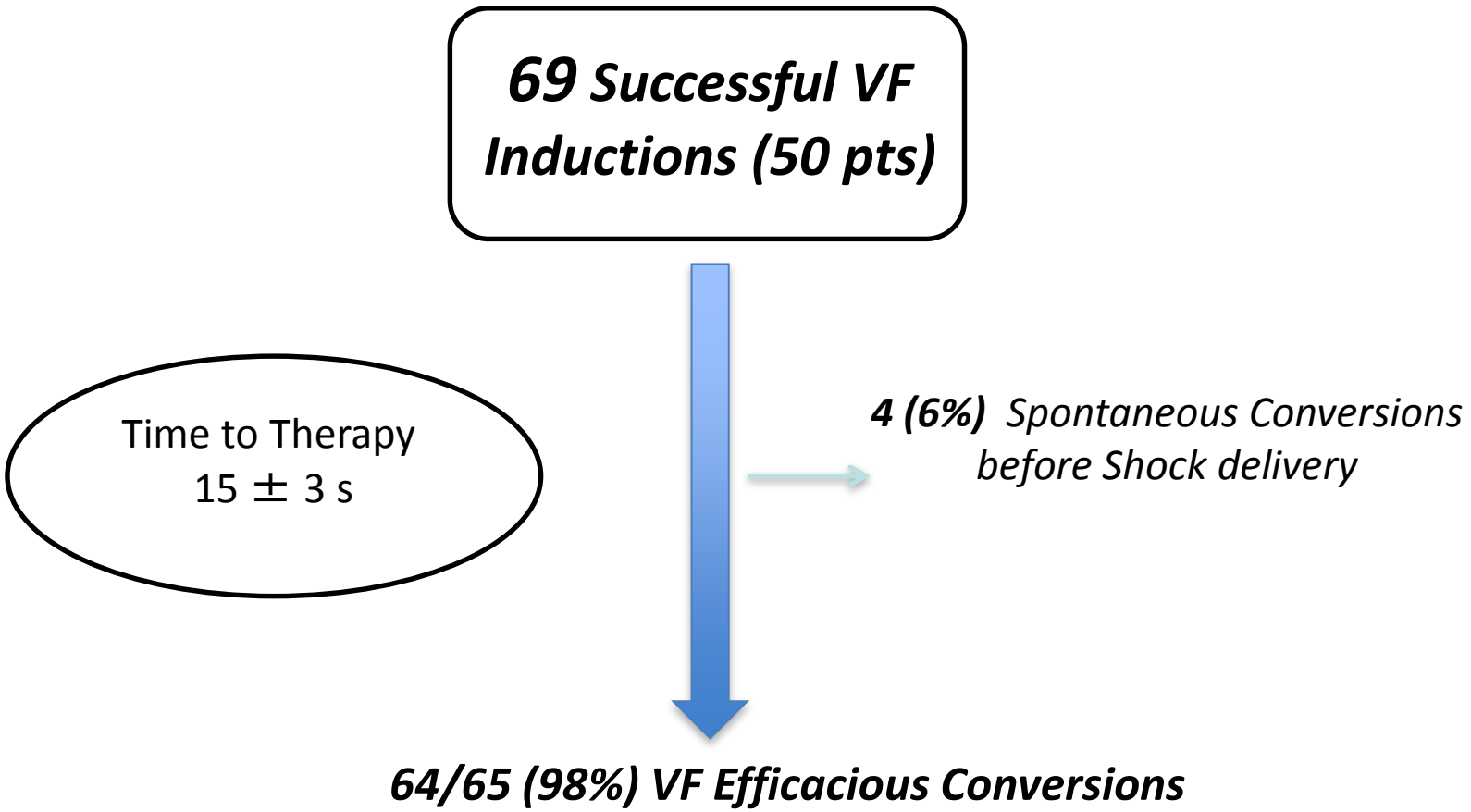


S-ICD may :

- avoid lead fracture and reduce sepsis management
- reduce inappropriate discharges due to SV arrhythmias
- improve LONG TERM RISK/BENEFIT RATIO + QOL
- increase acceptance by children and adolescents

S-ICD is not recommended when pacing is required

S-ICD DEFIBRILLATION TESTING EFFECTIVENESS REGISTRY



CONCLUSION

- In clinical practice individual risk assessment for SCD is easier if the ESC 2014 algorythm is used while discussing with the patient the available options for SCD prevention

- If the results of IDE and EFFORTLESS study are confirmed, S-ICD should be an option whenever pacing is not needed

Thanks !!

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